

THE SOCIAL IMPLICATIONS OF THALASSAEMIA<sup>MAJOR</sup> AMONG MUSLIMS OF  
PAKISTANI ORIGIN ~~IN ENGLAND~~-FAMILY EXPERIENCE AND SERVICE  
DELIVERY

by

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### ABSTRACT

This thesis is about the experience of Muslim British Pakistani families coping with thalassaemia (a chronic, inherited blood disorder) and the implications for service delivery. Its central concern is to illustrate that simplistic and culturally-biased assumptions are an unsatisfactory base on which to devise health service delivery for minority populations, and that with careful study it is possible to deliver culturally sensitive and appropriate services.

The thesis is written in four parts. The first part contains the research methods and the clinical aspects of thalassaemia. It also provides an introduction to the families in the study. The British Pakistani population is considered in the context of migration to Britain, which has created a plural society requiring adaptations to services to meet the diverse health needs of the different ethnic minorities.

The second part deals with the socio-economic and cultural background of British Pakistanis in Pakistan: this crucial to an understanding of their present situation. Family dynamics, marriage patterns and decision-making processes are explored, as is the central role of religion and kinship networks in the lives of British Pakistanis. It also examines their settlement process and present living conditions and illustrates how the social structures prevalent in Pakistan

have been re-established in England, albeit in a modified form.

The third part documents, using case studies, the experiences of British Pakistani families with thalassaemic children. These are analysed to highlight deficiencies in health service delivery and areas where cultural misconceptions exist. These areas require attention to provide an effective genetic counselling service for this population.

The final part examines the social and clinical implications of consanguineous marriage. It gives the results of a study showing increased frequency of consanguineous marriage among British Pakistanis than among Pakistanis in Pakistan. It then illustrates how kinship networks within communities practising this marriage pattern provide an opportunity to offer a genetic counselling service in a unique way, by making positive use of the practice. This proposed approach applies not only to thalassaemia but also to other inherited diseases.

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PREFACE

Some of the work presented in this thesis has been reported in the following publications:

Darr, A., Modell, B., (1988). The frequency of consanguineous marriage among British Pakistanis. Journal of Medical Genetics, Vol. 25, pp.186-190.

Petrou, M., Modell, B., Darr, A., Old, J., Kin, I., Weatherall, B., (in press). Antenatal diagnosis - How to deliver a comprehensive service in the United Kingdom. New York Academy of Sciences.

CONFIDENTIALITY

For the purpose of confidentiality, the cities in which the families in this study live have not been named. However, all the places referred to are areas, other than London, where large numbers of British Pakistanis have settled.

For the same reason changes have also been made to certain features in the family studies in chapter 12, which do not affect the authenticity of the studies.

STATEMENTS OF HEALTH PROFESSIONALS ABOUT BRITISH PAKISTANIS

"They leave everything to God and don't want to understand or use their initiative." - several health professionals.

"They shouldn't marry their cousins. If they didn't, they wouldn't have these diseases." - several health professionals.

"There's no point in offering them prenatal diagnosis. It's against their religion to terminate a pregnancy." - several doctors.

"They're uneducated people, you can't communicate with them." - a doctor.

"After a counselling session the parents are asked whether they have understood. Usually they have not, and have no idea of the importance of what was said." - a female doctor of Asian origin.

"Their children's health is not a priority with Pakistani parents, they're always taking them off to Pakistan." - a health visitor.

To an at-risk mother requesting prenatal diagnosis, "There is no such test for this condition." - a male G.P. of Pakistani origin.

"Why can't Pakistani parents make an effort and organise their own support groups, like the Cypriots." - a doctor.

PART ONE: INTRODUCTION

## RESEARCH BACKGROUND AND METHODS

### Origin of study

This research is a natural development arising from the need to deliver services for the treatment and prevention of thalassaemia to the British Pakistani population. The researcher was employed by University College, London, in the Department of Obstetrics and Gynaecology, but was based at the university in a city (city Z.) in England, where the fieldwork took place between September 1981 and June 1986. The supervisors were Dr. Bernadette Modell (B.M.), consultant in Perinatal Medicine at University College Hospital (U.C.H.) and Dr. Pamela Constantinides (P.C.), Honorary Research Fellow, London School of Hygiene and Tropical Medicine. For the purposes of supervision regular visits were made to London and B.M. visited city Z. where she had an honorary appointment at the university. The first year of the project was funded by a donation to the Department of Obstetrics and Gynaecology by Prince Sultan bin Abdul Aziz of Saudi Arabia, and the subsequent years funded by the D.H.S.S..

Thalassaemia was first identified as a problem among certain population groups in the U.K. in 1964 (Modell et al, 1972). Methods for effective management were developed at U.C.H. between 1964 and 1975 (Modell and Berdoukas, 1984, p.83). By 1973 the possibility of preventing the disease by population screening, genetic counselling and prenatal diagnosis first emerged (Kazazian et al, 1972) and between 1970 and 1980 a

genetic counselling service for thalassaemia had been developed and offered to the ethnic minority populations in North-East London at risk for the disease. In and around London British Cypriots are the population at highest risk though people of Indian, Pakistani, Italian and Chinese origin are also involved.

Contact between the U.C.H. centre and areas other than London where thalassaemia is mainly a problem for the British Pakistani population developed gradually. The first step was a referral in 1979 for prenatal diagnosis (P.N.D.) made by a haematologist in city Z. who recognized the importance of this problem locally. And almost uniquely among U.K. haematologists, this doctor had started screening and advising pregnant women of Asian origin. The policy was for all women in the ante-natal clinic to be tested; if a positive result was found the husband was also tested. At-risk couples so identified, were seen and advised by the haematologist. There were problems, however, as the screening was not always carried out and there were also unexpected problems in communicating the results and their meaning to some families. Nevertheless, a contact had been established with someone who perceived the need for a clinical service in the area, and realised the problems. For the recommendations of this research to be taken seriously and acted upon, the fact that there was local interest in the disease prior to the study was essential.

In 1979 B.M. was also contacted by a paediatrician from a small town six miles from city Z., regarding a muslim family of Pakistani origin with two thalassaemic children. The paediatrician had many problems communicating with the parents who were not complying with the recommended treatment for their children. There were many social and emotional problems in the family and the paediatrician was considering whether the children should be taken into care. B.M. responded by asking to meet the doctor and the family, together with any other families with thalassaemic children in his care (three families were involved), as she suspected families of Pakistani origin might encounter more problems with treatment and prevention of the disease, than the families of Cypriot origin in London. The meeting tended to confirm the suspicions. B.M. contacted the Regional Haematological Society and offered to meet and discuss the epidemiology, treatment and prevention of thalassaemia. The haematologist from city Z. was present at the meeting and it was proposed to establish an active collaboration on the problem. At the time, as an independant activity, B.M. sent a letter to all the paediatricians in the country through the British Paediatric Association seeking information about all the known cases of thalassaemia major in the country. This showed an increasing number of births of thalassaemic children to British Pakistani parents in parts of England with the highest concentration of this population and a relatively low "uptake rate" of prenatal diagnosis by British Pakistani couples



(Modell et al, 1985).

A number of questions requiring further study were emerging:

1. Do people of Pakistani origin have more difficulty than others in complying with the difficult treatment regime?
2. Does prenatal diagnosis have a low uptake rate
  - (a) because it is not wanted, or
  - (b) because there has been a failure in communication between the parents and the doctor and hence the parents are not aware of its availability?

B.M.'s own experience made her suspect that many problems were due to a failure in communication.

B.M. and the haematologist in city Z. agreed that it would be useful to carry out a study among the local families of Pakistani origin, especially as the local paediatricians were also concerned about the increasing number of thalassaemic patients, difficulties in communication and what they perceived as the parents' negative attitudes towards the management of the disease and prenatal diagnosis. Another provocation to carry out the study was the explicit view of some members of the medical profession that it was pointless discussing the option of prenatal diagnosis with muslims because "they do not agree with termination of pregnancy."

The established policy for offering a prevention service for

thalassaemia for most populations at-risk is to start an information programme for the community and offer prospective carrier detection and counselling, i.e., to detect carriers before they have had affected children. However, the W.H.O. recommended a 2-stage approach to establishing a screening service (Community control of hereditary anaemias, 1983). The programme should be started by working with couples who have already had an affected child, allowing problems to be identified and solved among families already acquainted with the disease and its implications; only when problems have been identified and resolved should one move onto carrier screening in the general population.

This recommendation, plus the events related above, seemed particularly relevant for studying a genetic prevention service in a population that appeared ambivalent towards the option of termination of pregnancy, and where the sensitive situation of a Muslim population with a consanguineous marriage pattern might also be an important issue. It was therefore decided to initiate a study of the implications of thalassaemia in British Pakistani families who already had an affected child.

It was clear to B.M. that technological improvements alone are insufficient for dealing with the disease comprehensively and a sociological/anthropological study of the problem was necessary to define appropriate approaches for the effective

delivery and utilisation of the medical service. Clearly ,the researcher had to be a woman of the same religious, cultural and linguistic background as the families and one who identified with her own culture and people. Because the research project entailed working with women in a population that practices segregation of the sexes and and because it involved discussion on personal issues such as childbirth, it was imperative that the researcher have enough maturity to discuss these and related issues. A research worker with the necessary attributes was found and an application for research funds was made.

#### Research methods

If one wishes to improve service provision, one logical step is to consult those affected by the service; the sample for the study was chosen according to this simple principle, that is, all the families with thalassaemic children in city Z.. It was not clear at this stage exactly how the research would develop but it was envisaged that the work would "snowball" and that the relevant issues would emerge.

To start the work the paediatricians with affected children in their care arranged a meeting between themselves, B.M. and myself (the researcher), gave permission to make the initial contact with the families and offered co-operation. They made the voluminous notes of all the patients available to B.M. for study and evaluation prior to meeting the families.

The first meeting with the parents and thalassaemic children took place at a clinical session in the Children's Hospital Outpatients Department, where B.M. and I together met families one at a time, before or after their encounter with their paediatrician. At the time, the children were under the care of different doctors. During the sessions B.M. assessed the children clinically and both of us asked the couples about their experience of thalassaemia, with myself interpreting when necessary. The couples were aware that B.M. was a specialist on their child's condition. I informed the couples that we wished to carry out a study to improve service provision and would like to visit them in their homes. All the families at the clinic consented to the visits.

After the sessions B.M. wrote a clinical report with recommendations for improved management of the disease. The implementation of the recommendations by the paediatricians resulted in increased haemoglobin levels maintained by transfusions and so in improved quality of life (Modell and Berdoukas, 1984). The study was specifically planned so that the introduction of the researcher would co-incide and be associated with improved treatment for the children.

It was clear from the beginning that the best approach to the study would be open participant observation. It would not have been possible to elicit the information required, nor to have gained access to the families' homes, without explaining

why the information was being sought. For a clear picture of changing attitudes, perceptions and responses to emerge over time it was necessary to participate to some degree in the families' lives for several years. The use of a questionnaire for eliciting information of a personal nature was inappropriate because questionnaires are impersonal and may be perceived as invasive if used at the beginning of a study such as this, creating a distance between the researcher and the interviewee. An open-ended questionnaire was used only at the end of the study.

Follow up visits began in October 1981, soon after the first meeting at the hospital whilst I was also familiarizing myself with the relevant clinical aspects of thalassaemia. As a study of this nature had not previously been conducted among British Pakistanis, I began these visits with scant knowledge of what would confront me. At this time, the views expressed by most health professionals were generally that: muslims are fatalistic and absolve themselves from making decisions by leaving everything to Allah (God); they do not believe in termination of pregnancy, it is very difficult to communicate with the families as most are uneducated and do not speak English, they do not turn up for appointments and so forth.

I had explained my role to the families during our first meeting at the hospital, but it was not understood by all the parents; their perceptions of me spanned a researcher, a

health worker, a doctor and an interpreter. I explained my role again but if unsuccessful I allowed their own perception of me to remain. Towards the end of the study virtually all the families understood my role as a researcher.

An appointment to visit was made by telephone when possible. In the absence of a telephone, as with three of the families, visits had to be unannounced. The regularity, frequency and content of the visits were determined by the particular issues in the family; visits to the home ranged from monthly to once a year on average, depending on the family and the intensity of the problems at the time. Each visit usually lasted at least one and a half hours. Discussions were loosely structured and centred around the thalassaemic child and the impact on the family, branching out into general discussions arising from those issues. Having begun with a discussion of the child, the parents had so much to relate that the cues for further discussion were taken from the parents.

Virtually from the first contact in the homes the meetings became counselling sessions. This was the first opportunity the parents had had to speak about their child and the disease with someone who spoke their language, gave them ample time and understood what they had to say. One of the most striking features during the initial visits was the isolation of the families due to their lack of awareness of the disease and of contact with other affected families.

The sessions were emotionally draining, particularly at first, as the parents related their distress, confusion, anger and frustration. They also asked many questions about the disease. Medical terminology, coined in English, had to be explained repeatedly in a different language and in a way which could be understood by them. Often their perceptions of the body and its workings differed from the western medical model. It was important not to discard the parents' perceptions of influences which may have caused the disease as nonsensical or irrelevant, but still to put forward an alternative explanation. Most of the information had to be repeated on many occasions, as it was difficult for parents to grasp the alien concepts I was trying to explain. Effective transmission of information was extremely difficult due to the lack of suitable health education materials.

I did not take notes during the initial visits in the homes but these were made immediately after the visit. I had taken notes at the first session in the Children's Hospital, and gradually resumed doing so in the presence of the parents, as the meetings became less emotionally charged and they grew more familiar with the fact that I was gathering information for a study to improve service provision, for others as well as themselves.

I was asked by parents about my family background, marital status and so forth, which I did not find intrusive. If I was

asking them personal questions it was natural that they would do the same with me. The dynamics of the interactions were essentially muslim and Pakistani in nature. The belonging to and understanding of Pakistani culture, and belief in Islam constituted a fundamental bond between myself and the couples. It conveyed non-verbally an implicit respect for, and understanding of the parent's beliefs, value system, way of thinking and rationalising their concerns. It also implied that we functioned within the same framework. Had I not been a muslim believer the gist and nature of the conversations would have been very different.

All the conversations were in Punjabi<sup>1</sup>, which is also my mother tongue. However, sometimes there was a mixture of Punjabi and English with those who were fluent in English. The majority of the families in the study were of rural origin and spoke a different dialect from myself. At first, when I began to speak, some families of rural origin noted that my Punjabi was urbanised and hence close to Urdu. Some began to

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<sup>1</sup> Punjabi is the mother tongue of those who originate from the Punjab and the adjoining areas of Azad Kashmir. The sociolinguistic history of this area is that most families in Britain with origins there speak varieties of Punjabi which are mutually intelligible. The local dialect of Punjabi is spoken in the home but Urdu is the language used for literacy.

Urdu is the national and official language of Pakistan and is used in Government offices and educational establishments. It is a language based on the speech of educated Muslims of Northern India and is written in the Perso-Arabic script. Among Pakistanis and British Pakistanis people who speak only Urdu are either migrants or refugees from India or educated, urban Pakistanis who have chosen not to speak their regional language in the home (Linguistic Minorities Project, pp.45-46, 1985).



Speak in Urdu but as I was aware this was not their mother tongue and they only spoke Urdu in response to my urbanised Punjabi, I continued to speak Punjabi. The conversation would then revert back to Punjabi, each speaking their own dialect. Rarely, the parents used words which I did not understand and I would ask what the words meant. This was treated as a learning process for me, as I was from a different region of the Punjab, rather than as a hindrance in communication.

It was important to conduct myself, and converse in a manner which emphasised the similarities between myself and the couples on the basis of religion, nationality and culture, rather than the differences in educational background and upbringing, as I have grown up and studied in Britain. I did not feel the need to wear traditional Pakistani clothes when in contact with the couples, although virtually all the women in the study wore them. I would sometimes dress in Western clothes and sometimes in Pakistani attire, dictated more by the weather than anything else. However, the clothes I wore always conformed to what would be termed respectable for someone of an urban background in Pakistani terms. I was always covered, except my hair, and the garments were loose-fitting in style. Had I, for example, worn a skirt with shoes rather than a long skirt with boots and not observed the nuances of etiquette, particularly in male-female behaviour, I feel I would not have gained as much entry into the lives of the families as I did.

I addressed the people I was talking to appropriately for their sex and age. If they were female and obviously younger than me I used their first name, if older than myself I addressed them as "baji" (elder sister) or "khala" (maternal aunt). I addressed males as "sahib" (Mr.). This established the relationships in a familiar, Pakistani context within which a certain respect and familiarity was accorded to each person. I was also referred to as baji, or in the feminine plural which accorded me respect and inclusion in their circle, whilst allowing a professional distance. The usage of terms such as "elder sister" are not taken literally when used by an outsider in Pakistani culture, but are used to express a desire to communicate in a familiar and trusting fashion rather than a formal one.

The isolation of the families made it clear that a family support group which would bring the families together was very necessary. When consulted, all the couples said they wished to meet with one another. Some had expressed a desire to do so prior to the consultation. The meetings, once started, were very successful, and also led to the establishment of a support group in a nearby city and stimulated interest in other cities. The first meeting of the group in city Z. was held in November 1982 and by the end of the research period six meetings had taken place. For the duration of the study the group meetings became an integral part of the families' involvement with the disease and improvement of its

management. Soon after the project started and the families were brought together, all the patients were brought under the care of one paediatrician and given standardized treatment. With the formation and success of the support group, the research snowballed from an analysis of individual families to include an analysis of the collective functioning of a group and the networks between the groups and families of different areas.

As well as taking on the role of counsellor I also found myself having to fulfill other functions such as arranging and running the support group meetings; making arrangements for children to visit other clinics for individual assessments, for example, for bone marrow transplants; arranging visits to London for prenatal diagnosis, sometimes accompanying parents on their first visits; and acting as an interpreter. This involved liaison and co-ordination with social workers, paediatricians and hospitals. The area of entitlement to state benefits was also touched upon.

The research showed very quickly that there were deficiencies in the service provided and that parents lacked, but wanted, information on the disease. To maintain a satisfactory relationship a researcher faced with gaps in service provision, is pulled willy-nilly into the situation of meeting the unmet needs. Although draining for the researcher, this situation is a precise indicator both of unmet needs and of how they can

be met, that is, the person of the researcher acts as a "social probe". The fact that I had to adopt the roles of social worker, counsellor, interpreter, advice worker and so forth, to carry on with the research, indicated the degree to which the roles were essential in providing an adequate service. In a similar manner the contact with other institutions also indicated where gaps in their service provision existed.

As envisaged, the research did in fact snowball. The study had begun by contacting families in the main study area (city Z.). Gradually, as the research progressed and people began to grow aware of the work, I was contacted by families in other cities asking me to visit or by health professionals asking me to visit families with thalassaemic children, give a talk or form a parent's association. Some of these families, along with others from the core group were partially studied and eleven of the total number of families were studied intensively. The amount of contact with families therefore varied. Families in the core study were more intensively studied than, for example, those the researcher travelled to see in a different city at a meeting arranged by a health professional. Even in the initial core group of families there were variations in the amount of contact and level of study, largely determined by the dynamics within the family facilitating or not facilitating discussion. In the absence of any teaching materials some families were more easily able

to grasp the information on the disease, due to their level of education, age and experience or because of a more desperate need to know, determined by the number of thalassaemic children in the family and their ages. A physical constraint on communication, which sometimes affected the information the families were able to give, was the space available in the home allowing privacy for a lengthy discussion. Certain topics were too personal to discuss if children or other family members also happened to be present. Sometimes the mother was able to create privacy, on other occasions she was not.

The research was by far richer for the increase in the number of families seen, even though some were seen only once for several hours or in a group. Impressions gleaned from the contact helped to validate certain findings from the core study group.

The research also snowballed in the area of support group formation. A health visitor in a nearby city, on hearing about the successful thalassaemia support group in city Z. approached me to form a support group which also ran successfully. During the course of the research these support group discussions were led by myself whilst the practical arrangements and follow-up visits were carried out by the health visitor. A further support group was set up by health professionals in a more distant city as a result of a talk

given to paediatricians and parents there. In 1988 I was invited to Denmark to present the findings of the research at a conference, as the same issues were emerging in Denmark as in England, with regard to their muslim populations from Pakistan and Turkey. There are now plans to start support groups for parents in Denmark also.

An added and important dimension of the work was that I chose to become involved in local health issues in city Z.. I wished to place the experience of thalassaemia in the broad context of other health issues, particularly those facing the ethnic minority population in the city. Over the years I was Chairperson of the local Community Relations Council (C.R.C.) Health Initiative, on the C.R.C. Health and Welfare Panel, on the management committee of the Asian Women and Girls' Resource Centre, a member of the Asian Workers Support Group and involved in the campaign for a Well Woman Clinic. I perceived that some of the issues relating to inadequate provision for thalassaemia also related to other areas of health care provision for ethnic minorities. Through this involvement I was able to discuss my views with other health workers and to put thalassaemia on the agenda as a health issue requiring attention at a local level.

In 1986 I visited the Thalassaemia Centre in Karachi, Pakistan for ten days at the invitation of the Fatimid Foundation, with two members of the Perinatal Centre staff. During this period

I interviewed parents of 128 thalassaemic children who were registered with the Foundation<sup>2</sup>. I stayed on in Pakistan for a further two and a half weeks, travelling around Karachi and the Punjab. This visit plus three subsequent holidays in Pakistan have allowed an understanding of the issues facing families with thalassaemic children and an observation of life in Pakistan with regard to some of the issues emerging in the research.

It was important that this study was able to get funding for several (nearly five) years. As the study was about analysing the total experience of the disease and its impact, this allowed an analysis of change over time, including changes in attitudes due to developments in personal situations or developments in technology. The time factor allowed greater depth and insight and hence a clearer picture emerged.

In the last months of the research an open ended questionnaire was used to fill in the gaps in the information collected with the intensively and partially studied families. Due to the informal relationship which had developed with virtually all the families, going through the questionnaire sometimes took a whole day, with general socialising in between. Where the families had moved home during the research period, visits

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<sup>2</sup> This work is reported in Modell, B., Petrou, M., Darr, A., (1986). Thalassaemia in Pakistan. (unpublished report), but does not form part of this thesis.

were made to their new homes. The final visits to the families were made during the period April to June 1986, but further contact has been made with some families by telephone or visits since then.

Questions such as whether the family owned a car, telephone and so on were not included in the questionnaire so as to limit the questions to those which were, from the parents point of view, obviously relevant to the study. However, I noted such socially relevant factors by direct observation.

When the nature of the families' situation became apparent I made a particular effort to feed back information to the families and to take on a developmental role as an intrinsic part of the work, for example, by the establishment and running of the support groups, as I was aware of some resentment among community workers and residents in the city who felt that a lot of research had been carried out among British Pakistanis without their seeing the benefits of that research.

This research then is based on an exploration in the family setting, focussing on the child and the family and then encompassing the families as groups. The involvement in local community and health organisations allowed an understanding of inequalities in health care delivery, and its pervasiveness in areas other than genetic health services. This latter



experience is not the subject of this thesis but it consolidated my perceptions of the role and impact of inequalities in healthcare provision and instilled a confidence in the findings of this research and the need for change. The racial politics of health provision to ethnic minorities is well documented elsewhere (Brent CHC, 1981, Pearson, 1986, Sivanandan 1976, Townsend, 1988, Torkington, 1983).

Although, within the families, the research progressed smoothly there were unforeseen problems centring around the research team's relationship with the doctors in city Z., who at the beginning of the study had been very co-operative and helpful, but who felt some hostility towards us at the end of the project. During the course of the project it was difficult to pinpoint precisely how and when the relationship deteriorated but the situation has been much easier to analyse with hindsight. The conclusions we have now reached may prove useful for other similar research projects in avoiding unnecessary pitfalls. However, the creation of hostility may, we feel, be an inevitable part of research into unmet needs.

Having established the project and the researcher in city Z., B.M. returned to continue her work at U.C.H. in London. B.M. had expected that I would maintain close links with the haematologist, in whose department I worked alone for a few months, collecting data at the beginning of the study.

However, when this period was over I was based at the local University, began meeting the families and had little need to contact the doctors. There were just a few occasions of contact, either when I visited a family at the request of one of the doctors, and at joint meetings with the haematologist when B.M. visited city Z. These meetings, at which the project findings and the families were discussed amongst other things, became less frequent as the work progressed, due to B.M.'s commitments with regard to the development of chorionic villus sampling (see below). Distance made it difficult for her to pick up the warning signs. My interaction with the doctors had not been formally stated by the research team or by the doctors and therefore remained unstructured.

During 1982 there was a breakthrough in developing procedures for chorionic villus sampling at U.C.H. which became B.M.'s principle preoccupation, as this was a major development in the field, with particular relevance for Muslim couples. Hence, at this time B.M.'s visits to city Z. became infrequent and I travelled more to London for supervision. At the time, my work consisted mainly of visits to the families, the support groups, giving talks and involvement in local health organisations. As the work was progressing well I did not feel the need for contact with the doctors and I had not perceived any expectation on B.M.'s part of ongoing contact. B.M. at one point had asked me to prepare case studies on the families for the newly appointed paediatrician who was to care

for all the children in city Z., but at the time, with only hand-written notes I found this a formidable task, was busy with other work and did not realise the importance of providing this information, if only as feedback of the project.

Within this context, another unwitting and major mistake was made; I presented the tentative findings of the research at a national conference of paediatricians with B.M.'s approval but without consultation with the local doctors. Although the talk was prefaced by saying that the children were clinically extremely well looked after, it then went on to point out the deficiencies of the service provided and the stereotypical assumptions that were made about the families behavioural responses. One of the paediatricians from city Z. was present at the conference and was quite rightly offended at hearing publicly, for the first time, what may have been better discussed beforehand in private. I was subsequently invited by the paediatrician to present the same talk to the doctors in city Z.. At the talk I felt some covert hostility from some, though not all of the doctors.

As the deteriorating situation became more apparent apologies were offered and efforts made to make amends. However, the research was coming to a natural conclusion by this time and my withdrawal from city Z. marked the termination of any active interaction.

There were several reasons, we feel, for these problems. There should have been a formal timetable established at the beginning of the research to give continual feedback to the doctors on the progress of the research and the emerging issues as an integral part of the project. We recognised that feedback to the families was important but failed to do so in the case of the doctors.

However, we feel there were also other subliminal dynamics relating to power. The medical profession is an institution vested with power by society and which, by its exclusivity, nurtures feelings of self sufficiency among its members, who remain unused to criticism, particularly from outsiders. In the case of this research the outsiders were on a number of levels: a team from outside of city Z., from the power base of London to the provincial North; a sociologist informing the medical profession of its deficiencies.

When researching in the area of unmet needs a certain degree of hostility is inevitable as deficiencies in service delivery are highlighted and service providers are forced to assess their roles. The person or group pointing out the deficiencies is considered to be "abrasive". However, it is also recognised that a degree of abrasiveness is constructive and inevitable for adaptation and change. This process can be made easier by frequent contact between the researcher and the

service providers, allowing the latter to be in touch with the need for change as a gradual process.

## GENETIC COUNSELLING

The following are authoritative and established views on genetic counselling as a process:

"Genetic counselling is the process by which patients or relatives at risk of a disorder that may be hereditary are advised of the consequences of the disorder, the probability of developing and transmitting it and of the ways in which this may be prevented or ameliorated (Harper, 1988, p.3)."

"Although counselling should be non-directive, this does not simply mean telling people the facts and leaving them to make their own decision. Counselling is a special skill that depends on training and the ability to communicate, and it involves actively helping couples to reach decisions in the context of their unique medical, moral and social situation (The Royal College of Physicians, 1989, p.31)."

The first clinic for genetic counselling in the U.K. was developed at the Hospital for Sick Children at Great Ormond St., London in 1946. Although there have been changes since then in terms of social attitudes and carrier detection plus prenatal diagnosis of affected fetuses, the development of a community genetics service is still in its infancy. The recently published report of the Royal College of Physicians (1989), entitled "Prenatal Diagnosis and Genetic Screening-Community and Service Implications" is the first attempt to devise a national approach for the delivery of a community genetics service to the whole population. However, there is a functioning structure of clinical genetics and genetic counselling services in operation for inherited diseases; provision for haemophilia and phenylketonuria which largely

affect the majority population is excellent (Prashar et al, 1985, p.51) whereas services for thalassaemia and sickle cell disease, which mainly affect ethnic minorities, remain inadequate (The Royal College of Physicians ,1989, p.26).

## THE CLINICAL ASPECTS OF THALASSAEMIA

The following chapter draws largely on Modell and Berdoukas' comprehensive book, "The Clinical Approach to Thalassaemia".

### The thalassaemias

The thalassaemias are inherited disorders of the haemoglobin of the blood (haemoglobinopathies). They are inherited as autosomal recessives (see below). "The haemoglobinopathies" is a collective term for such inherited disorders, the commonest being thalassaemia and sickle cell disease. There are several types of thalassaemia, the most common and most severe of which is  $\beta$  (beta) thalassaemia major (also referred to as Cooley's anaemia or Mediterranean anaemia). The vast majority of the children in this study suffered from this form of the disease.

### Thalassaemia intermedia and other forms of thalassaemia

Thalassaemia intermedia is usually understood to mean thalassaemia of a milder kind. Some patients have minimal disability whilst others are chronic invalids. Milder syndromes of thalassaemia occur wherever thalassaemia is common. Other less common forms of the disease include HbS/thalassaemia, HbE/thalassaemia, delta-beta thalassaemia, Haemoglobin Lepore and alpha thalassaemia.



### Thalassaemia trait

Thalassaemia trait is also referred to as thalassaemia minor, the heterozygous form of thalassaemia, or healthy carrier of thalassaemia. It is not a disease but a characteristic of the blood which is inherited like eye, hair or skin colour. The thalassaemia gene is carried as a single dose on one of the autosomes and is found in both males and females.

Because of the multiplicity of terms to describe the same disease and since most of the children in this study suffered from thalassaemia major, henceforth in the text, the word thalassaemia will be used to refer to  $\beta$  thalassaemia major and thalassaemia trait to refer to the heterozygous form of the same disease. Thalassaemia intermedia will be used when indicated. "The thalassaemias" is used to refer collectively to all the forms of thalassaemia.

Before embarking on a description of the clinical aspects of thalassaemia it is important to describe their autosomal recessive inheritance and the molecular basis of thalassaemia.

### Genetic inheritance

Thalassaemia is among the inherited diseases that are caused in a relatively simple way by defects of single genes. Genes are carried on the chromosomes, of which there are twenty three pairs in humans: twenty two pairs of autosomes and two pairs of sex chromosomes, the X and Y chromosomes (fig 1.1c).

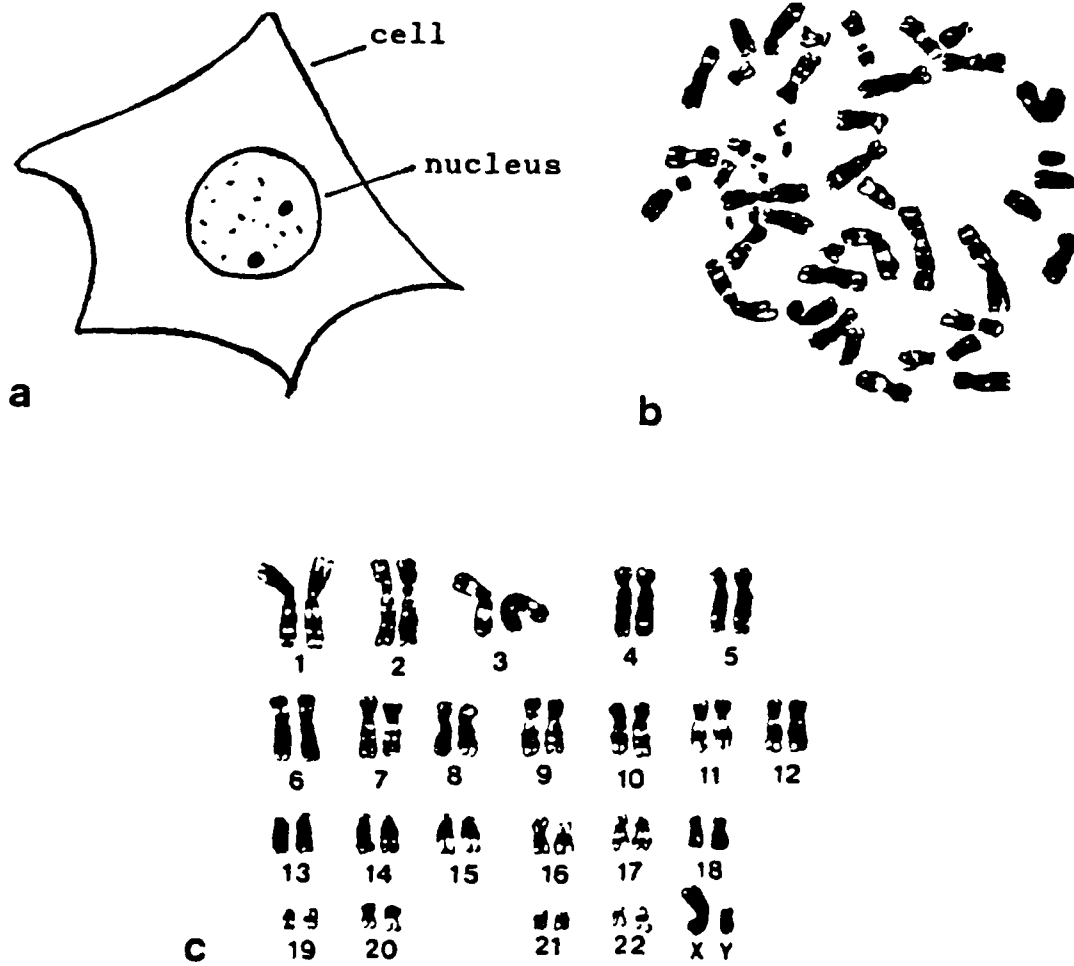


Fig. 1.1: Human chromosomes

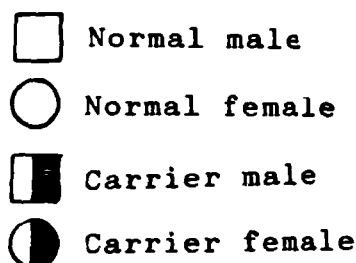
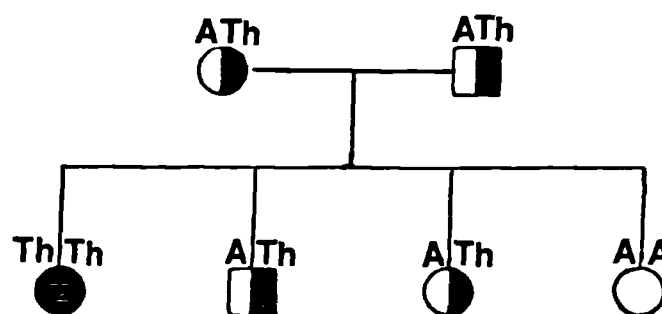
The nucleus of each cell of the body (fig. 1.1a) contains the chromosomes which are long strands of D.N.A. coated with proteins. The D.N.A. carries the "genetic code" for all the constituents of the developing embryo and the adult. When cells divide the chromosomes separate out, and can be studied microscopically (fig. 1.1b). If special stains are used they take on a banded appearance. Each of the twenty three pairs has a characteristic banding pattern. To examine them more closely, a photograph is taken, and the chromosomes are cut out and arranged in pairs (fig. 1.1c).

One of each pair of chromosomes is inherited from the mother and one from the father. Females have two X chromosomes and males have one X and one Y chromosome. The inheritance of genetic traits differs depending on whether they are carried on an autosome or on the X chromosome (very few traits are carried on the Y chromosome). When both the genes for a given characteristic on a chromosome pair are identical, the person is "homozygous" for that characteristic. When one is a variant gene the person is heterozygous for that characteristic.

There are four different patterns of inheritance: autosomal dominant and autosomal recessive, X-linked recessive, X-linked dominant. Since the haemoglobinopathies are examples of autosomal recessive inheritance, this is the only pattern that needs to be described here.

### Autosomal recessive inheritance

Genes that are inherited as autosomal recessives are located on one or other of the autosomes. They can therefore show effects in both males and females, but only when an individual has inherited a double dose of the gene, one from the mother and one from the father, i.e., he or she is homozygous for that condition. Autosomal recessive inheritance using thalassaemia as an example, is shown in fig. 1.2.



A Gene for normal haemoglobin  
 (haemoglobin A)  
 Th Gene for thalassaemia

Fig 1.2: Autosomal recessive inheritance of thalassaemia.

### Biochemical basis of thalassaemia

Adult haemoglobin (HbA) is a molecule consisting of two  $\alpha$  (alpha) and two  $\beta$  (beta) globin chains. The basic defect causing thalassaemia is an imbalance in the synthesis of  $\alpha$  and  $\beta$  chains. The synthesis of  $\alpha$  and  $\beta$  chains for normal red

cells, thalassaemia trait red cells and thalassaemia major red cells is shown in figs. 1.3, 1.4 and 1.5.

There are 30 pgs of HbA in a normal red cell. HbA<sub>2</sub> (formed in smaller quantities by the synthesis of  $\alpha$  and  $\delta$  (delta) chains is less than 3%. The synthesis of the  $\alpha$ ,  $\beta$  and  $\delta$  chains is directed by the corresponding genes. In normal red cells equal amounts of  $\alpha$  and  $\beta$  chains are synthesised.

In thalassaemia trait one  $\beta$  gene is not active, so only about half the normal amount of  $\beta$  chains are made. However, the cell finally contains about 20pg of HbA rather than 15pg as would be expected as there is some "compensation" within the cell. The bone marrow also compensates for the fact that each cell contains less than the normal amount of haemoglobin by making twice as many red cells, so there is little or no anaemia in thalassaemia trait. The HbA<sub>2</sub> is higher than 3.5% which is very important for the diagnosis of carriers.

In homomozygous thalassaemia no adult haemoglobin is made as no  $\beta$  genes are active. At birth there are no symptoms because of the presence of about 80% fetal haemoglobin in the normal neonate. As the fetal haemoglobin declines, adult HbA synthesis cannot take place, due to the inactivity of the  $\beta$  genes. The failure of HbA synthesis causes severe anaemia. As the red cells are abnormal and quickly destroyed the bone marrow becomes very active in order to compensate for the anaemia. If treatment with blood transfusions is not started

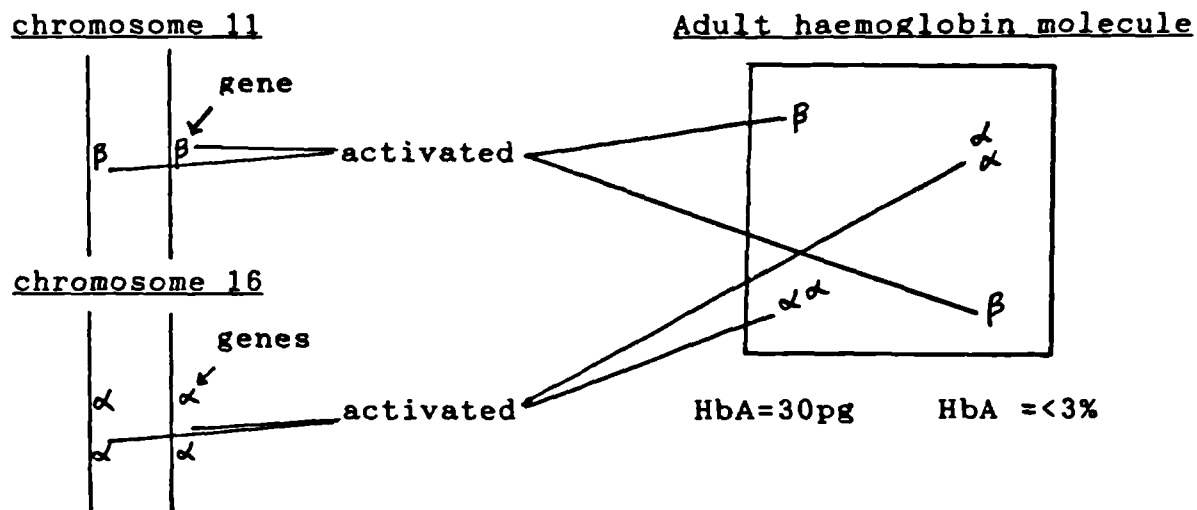


Fig 1.3: Normal  $\alpha$  and  $\beta$  chain synthesis.

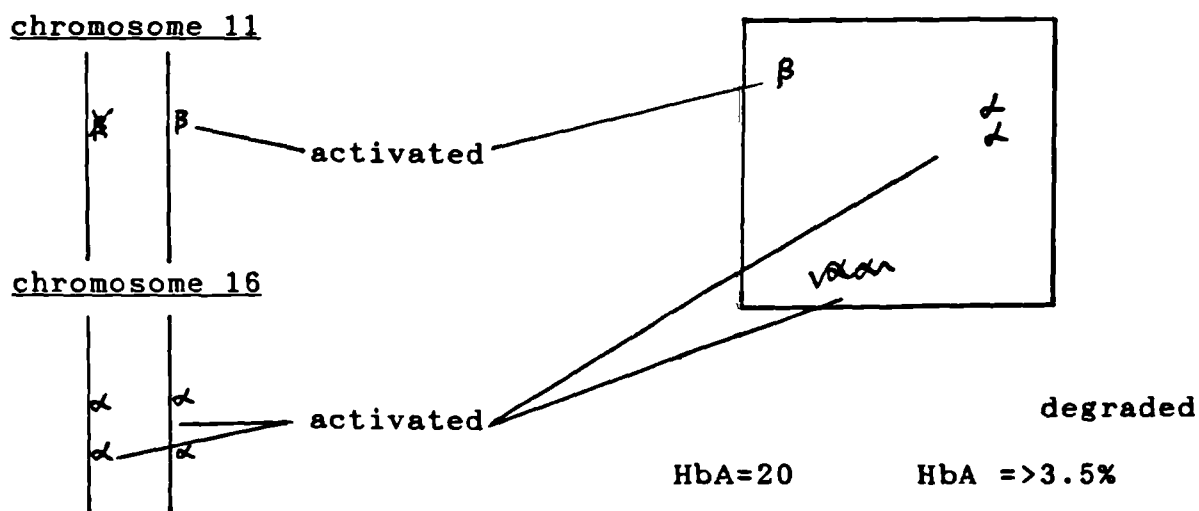


Fig 1.4:  $\alpha$  and  $\beta$  chain synthesis in thalassaemia major trait.

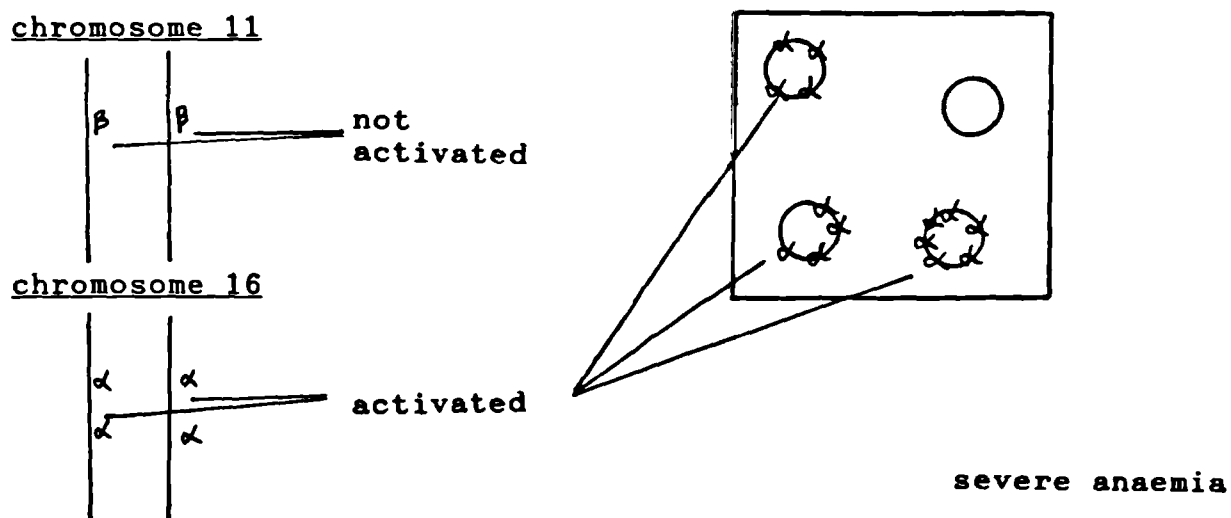


Fig. 1.5:  $\alpha$  and  $\beta$  chain synthesis in thalassaemia major.

early the result can be skeletal malformations (Petrou, 1987; Colvin, 1977).

### Carrier detection

In most recessively inherited diseases there are no tests that can identify carriers unambiguously. In these cases the only way a couple of carriers can find they are at risk for a particular disease is by actually having an affected child and getting it diagnosed. Usually affected children are born without a family history of the disease. The sporadic nature of recessive disease needs quite a lot of understanding.

Thalassaemia trait, however, can be detected simply, cheaply and reliably. The procedure involves taking a small blood sample which is analysed using standard haematological techniques. The results are made available within a week. So, in principle, screening can be offered and at-risk couples informed of their risk prior to reproduction.

### The implications of having thalassaemia trait

A person having thalassaemia trait, i.e., a carrier of thalassaemia, is usually absolutely normal, though some may be very slightly anaemic. Thalassaemia trait is often brought to light when an individual has a blood test for a completely unconnected reason or sometimes only after they have produced a child with thalassaemia major. The chance of passing on thalassaemia trait when one partner has

thalassaemia trait and the other has normal blood are shown in fig. 1.6. There is a 50% chance in each pregnancy that the child will have thalassaemia trait. The risks connected to each pregnancy if both partners have thalassaemia trait (an "at-risk couple") are shown in fig 1.7. In each pregnancy there is a 25% chance of having a child with thalassaemia major, a 50% chance of having a child with thalassaemia trait and a 25% chance of having a normal child.

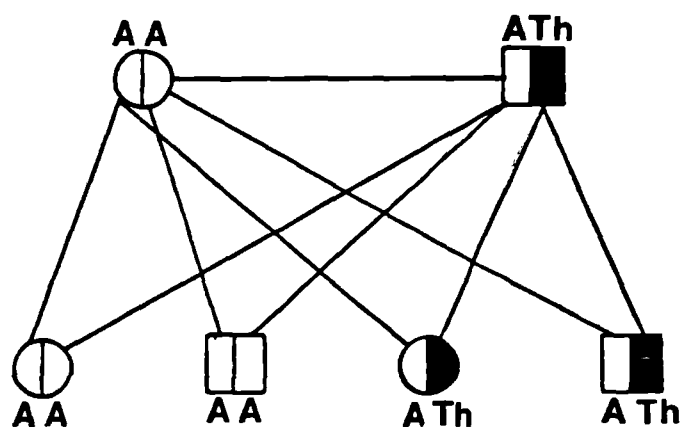


Fig 1.6: Risk of passing on thalassaemia trait when one partner has normal blood and the other has thalassaemia trait.

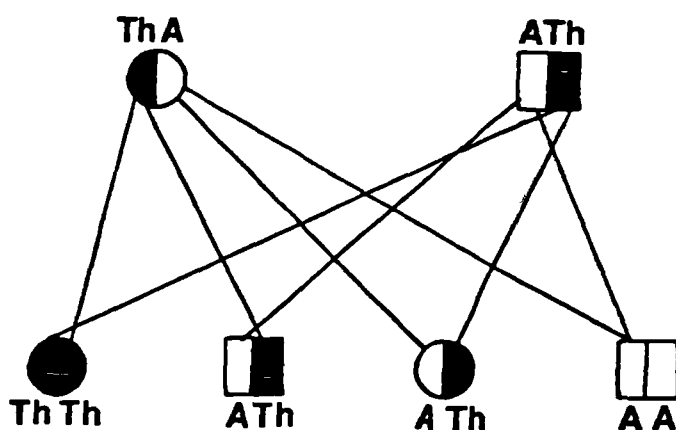


Fig 1.7: Risks connected to a pregnancy if both partners have thalassaemia trait.



Reproductive options for a couple both carrying thalassaemia trait

1. To separate and reproduce with partners with normal blood.
2. To marry and:
  - a) Not reproduce, by using contraception (would require termination of accidental pregnancies).
  - b) Reproduce and risk affected pregnancies without making use of treatment.
  - c) Reproduce and proceed with treatment of any thalassaemic children.
  - d) Reproduce using prenatal diagnosis to detect affected fetuses in time for the option of termination of pregnancy.
  - e) Reproduce normally, refuse prenatal diagnosis and option of termination of affected pregnancies, with possibility of bone marrow transplantation for any affected children.

Thalassaemia major without treatment

Dr Alan Fawdry's account of the condition of thalassaemia sufferers in Cyprus in 1944 provides an illustration of a child's life without treatment:

"The usual history is that the child was apparently quite healthy for the first few months of life and then fell ill with irregular fever and intestinal upset, following or during which pallor and weakness made their appearance and remained permanent. However, the onset is rare before three months and extremely uncommon after eight years.

Accompanying the anaemia are many other signs of disease, of which enlargement of the spleen and changes in the shape of the bones of the head, giving a mongoloid face, are the two most striking features. The actual disability of the child depends entirely on the degree of anaemia; if the concentration of haemoglobin in the blood is less than 25% of normal the child is usually unable to walk or play owing to the weakness and shortness of breath accompanying every muscular effort; between 25% and 50% the child can walk and play but can scarcely run, and when over 50% the disability is very slight. No pain is caused and the child's mental development is normal, in fact a fair proportion seem to be above the average intelligence.

The course of the disease is variable; in general the earlier the onset, the more rapid the development and the greater the degree of the anaemia. After the period of the febrile onset has passed the body usually seems to adapt itself to the new conditions of the blood and the progress of the disease is arrested for a period of years. Gradually, however, the child's demands on its body exceed the latter's capabilities and heart failure sets in, though the child's death is usually precipitated by some intercurrent infection, e.g., bronchopneumonia.

Thus ends a short span of life, rarely more than 8, often less than three years, which brings about as much misery to the parents as to the suffering child.

In a few cases even seriously anaemic children do survive to puberty and beyond but their existence is often rather miserable and they have not the strength needed to earn a living for themselves. Others, it is at present impossible to estimate how many, develop a mild degree of anaemia with enlargement of the spleen and suffer only slight weakness which does not hinder them in their work and everyday life."

Although treatment for thalassaemia in Western Europe has improved significantly in the past few decades Fawdry's description of a thalassaemic child's life still holds true for most of those born in Pakistan and other economically developing countries today.

### Treatment of thalassaemia

Children with thalassaemia major usually present between three months and two years of age with pallor, loss of appetite, vomiting, failure to gain weight, sleeplessness and irritability. The haematological findings in an affected child are severe anaemia with grossly abnormal red cells, and especially nucleated red cells in the blood film, raised HbF (fetal haemoglobin), and the diagnosis of thalassaemia trait in both parents.

Transfusions are the mainstay of treatment and are needed for life at 4-6 weekly intervals. For the child and a parent every transfusion can mean three visits to the hospital. The first and third visits are made to check the haemoglobin levels from blood samples taken by a finger-prick whilst the second visit is for the actual transfusion which usually entails an overnight stay.

Each unit of blood contains 20mg of iron (Fe). A patient usually requires about 20 transfused units of blood per annum, resulting in an annual intake of about 5 grammes of iron. Iron cannot be disposed of by the body, therefore, it must be stored. A normal adult body stores only one gramme of iron, so patients in fact store several times the normal amount of iron annually. As iron is toxic, when a patient's storage capacity is exhausted complications arise. This usually occurs after about twelve years of transfusions. The

most important complications are heart failure, failure of growth, puberty and sexual development, and endocrine problems, such as diabetes and hypothyroidism. These problems can be prevented and are controlled by the intensive use of an iron-chelating drug desferrioxamine, also referred to as Desferal. Desferal is administered most effectively by subcutaneous infusion using a syringe driver (fig 1.8). The process of administering the Desferal involves the parents or patient mixing the required amount of distilled water and Desferal into a syringe, which is attached to the small portable syringe driver (commonly referred to as "the pump"). A butterfly needle with a one-metre-long tube attached to the syringe is inserted into the abdomen and secured with a plaster. The pump is attached to the body in a holster or placed beside or under the bed all night. The infusion takes approximately eight hours and the procedure is repeated five to seven times a week. Prior to the introduction of the syringe driver, Desferal was administered by the less effective method of intramuscular injections.

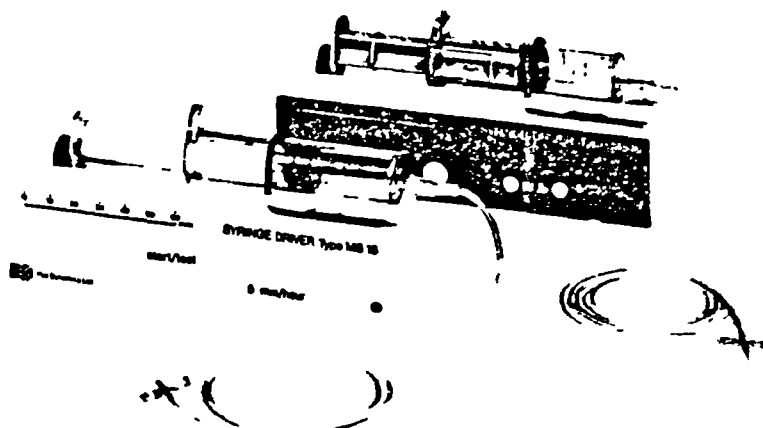


Fig 1.8: Portable syringe driver.

The survival of thalassaemic children in relation to treatment is shown in fig 1.9. At present the prognosis for a well-treated patient is considered to be "open-ended, i.e., unknown but optimistic.

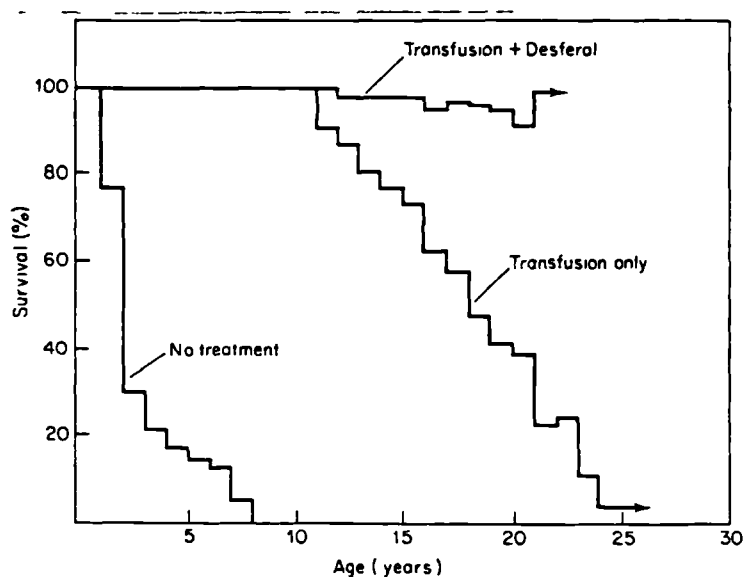


Fig. 1.9<sup>1</sup>: Survival in thalassaemia in relation to treatment.

The treatment for thalassaemia is aggressive, demanding and burdensome, and forces adjustments in the daily life of the family. Employment is affected and loss of earnings incurred by parents needing to take time off work to take children to hospital. Parents and children alike are often distressed by the frequency with which needles need to be inserted into the body, particularly if the first attempt at insertion is

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<sup>1</sup> Source: Modell and Berdoukas (1984), p.171.

unsuccessful and repeated attempts need to be made. A child's veins need to be treated with care because if they become inflamed they can become blocked and another vein must be found. Often other children and relaxation time are neglected on account of the amount of energy and emotion channelled towards the sick child.

#### Bone marrow transplantation

Presently, a successful bone marrow transplant is the only complete cure for thalassaemia major. The operation involves the suppression of the affected child's bone marrow using drugs, so that the bone marrow of a compatible donor can be grafted. The basic requirements are a child as young as possible and a fully compatible donor (usually a sibling), who may or may not carry thalassaemia trait.

Blood samples are taken from parents and siblings for H.L.A. tissue typing. If a compatible donor is found the family attends the transplant centre for further confirmation of tests and to be informed of the procedure. Prior to the procedure, the child is required to stay in a sterilised room isolated from other children, with one parent who must wear a gown and mask to prevent infections. The hospital stay usually lasts about three to six weeks. Drugs are administered to eliminate the child's own bone marrow, which leaves the child open to infection and also causes the child's hair to fall out subsequently. A few days later

marrow is taken from the donor using a needle, under general anaesthetic. One or two bones, normally the hip bone, are used as the source. The marrow is processed and then infused into the recipient. If the procedure is successful the recipient starts to produce red cells, white cells and platelets during the next three to six weeks. Throughout this time the child is highly susceptible to infection as the child's own defence mechanism has been suppressed, so everything he/she comes into contact with must be sterilised. Even after a transplant has been successful care must still be taken, as the child's immune system remains below normal for some time. A complication of the operation is that the graft may not take at all, or graft versus host disease (G.V.H.), a chronic condition, may set in. Four children in this study underwent the procedure. Two are now healthy and two died.

Bone marrow transplantation is particularly relevant to populations like the Pakistanis that practice consanguineous marriage and have large families, as both these factors increase the chance of finding a compatible donor. It may even be viewed as an alternative to terminating an affected pregnancy.

The centres of expertise for bone marrow transplantation are in London and hence for families in this study the operation entailed many journeys back and forth. Parents need to take

time off work, losing earnings, make arrangements for their other children to be taken care of and pay the cost of travel, unless they are unemployed, in which case travel warrants need to be applied for. Any delay in processing an application can create panic for fear of missing an appointment.

For mothers in this study the hospital stay was also an alienating experience. Unable to speak English fluently the weeks in hospital can be lonely, with telephone conversations being the only contact with their spouse and other children. Any extra journeys made by family to visit entail extra expense for the journey as well as overnight accommodation. An added complication is having to purchase meals and eat an English diet, which can often seem unpalatable for someone not used to it, the only option being to find and go to a Pakistani restaurant, something none of the mothers had ever done before. For most women the whole experience of living away from home and venturing out into a city alone may be their first.

#### Growth and puberty

In general, growth in thalassaemic children progresses normally to eleven years, but unless they have been on Desferal regularly from early in life there is usually an abrupt slowing to about half the normal rate between the eleventh and twelfth year. Many patients experience



difficulty with sexual development, varying degrees of stunting and emotional stress. Those who have received Desferal regularly from about seven years of age often, but not always, have better growth and development. As Modell and Berdoukas succinctly state:

"It is impossible to exaggerate the importance to the patients of growth retardation and delay of puberty. It brings their abnormality home to even the most resilient individuals. They feel inadequate and threatened and may become very depressed, but they are often too shy to discuss the topic with parents or their doctor, and all too often the physician treating them fails to raise the subject out of delicacy or unfamiliarity with the physical and mental problems of adolescents (pp.193-4)."

#### Main causes of death

The main factors that can lead to death in thalassaemia major fall into two groups: facets of the disease (anaemia, hypersplenism) and complications of treatment (accidents of transfusion, post-splenectomy infections, iron overload). The causes of death from thalassaemia in Britain between 1955 and 1966 are shown in table 1.1.

<u>Cause of death</u>	<u>No. of patients</u>
Anaemia	17
Bleeding due to thrombocytopenia	4
Infections	10
Complications of transfusion	1
Iron overload	57

Table 1.1<sup>2</sup>: Causes of death from thalassaemia in Britain.

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<sup>2</sup> Source: Modell and Berdoukas (1984), p.152.

### The future

"Since 1964 there has been no fundamental change in the instruments available for management of thalassaemia, but in the intervening years we have learnt how to use regular blood transfusion and parenteral administration of desferrioxamine to their best advantage. As a result, thalassaemia major has been transformed from a disease with a uniformly depressing outlook, to a manageable condition with a good prognosis for survival, work, marriage and reproduction (Modell and Berdoukas, 1984, p.388)."

Improvements in treatment for the future are expected to be the development of an oral iron chelator to replace the burdensome use of the pump; in utero bone marrow transplantation; and gene therapy by influencing the "fetal switch". The fetal switch occurs at about thirty five weeks gestation when the  $\alpha$  genes are switched off and the  $\beta$  genes start to be activated, so that HbA begins to replace HbF in the circulation. If the process could be prevented or reversed patients with thalassaemia could be effectively cured as they usually have no disorder of the  $\alpha$  genes. Modell and Berdoukas expect that a cure for thalassaemia will be available by the year 2,000.

### Prevention of thalassaemia

Fetal diagnosis and selective termination of pregnancy as a preventative measure for thalassaemia has been available only since 1977. An at-risk pregnancy can be tested for thalassaemia by two methods; fetal blood sampling (F.B.S.) and chorionic villus sampling (C.V.S.).

### Fetal blood sampling

Fetal blood sampling is performed at 18-20 weeks after the last menstrual period. A thin needle is inserted into the womb under ultrasound guidance, a small blood sample is taken from the fetus' umbilical cord. The needle does not touch the fetus itself (fig. 1.10). The mother is asked to attend the hospital a few days before the examination for an ultrasound scan. This provides a televised picture and allows an accurate assessment of the duration of the pregnancy. The scan is painless and almost certainly harmless. On the morning of the test the mother may be given a sedative to help her to relax and to make the baby lie still. A local anaesthetic is injected on the point of entry on the abdomen. The mother experiences a momentary feeling of pressure when the instrument is inserted and a rather sharp pain as it passes through the uterus wall. Apart from this, she should feel no pain. The test takes approximately twenty minutes and there are practically no risks to the mother undergoing it. After the test the mother may rest for some hours in hospital.

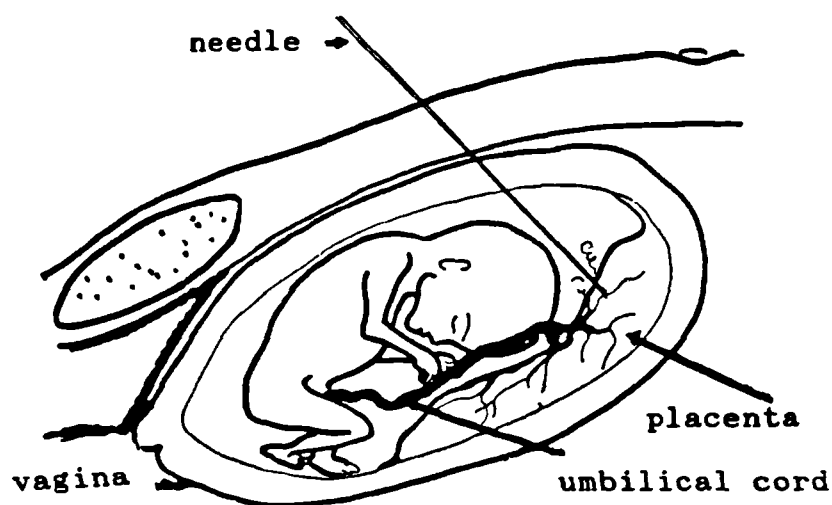


Fig 1.10: 18-week fetus and membranes, about half natural size; showing fetal blood sampling.

The results of the test are available in two to four days. The uterus may become irritable after the procedure, causing contractions which can usually be stopped with treatment. The risk of a miscarriage following modern fetal blood sampling is thought to be 1-2%. However, at the time this study was done it was higher, at 2-4%. To minimise these risks the mother is advised after the test to avoid carrying heavy objects and tiring housework and to refrain from sexual intercourse for one week.

The laboratory tests are complicated, and great care is taken. However, there are pitfalls which can lead to a mistaken diagnosis and this has occurred in all the centres of the world. The risk for such an error is calculated to be

1%. If, as sometimes happens, the results are doubtful, the test may have to be repeated two weeks later.

If the test shows that the baby has thalassaemia major, a termination of the pregnancy can be carried out, if requested by the parents. Termination at this stage is carried out by injecting prostaglandins into the uterus. These commence labour pains and start a miscarriage. An outdated way to stop a pregnancy after eighteen weeks is by a form of caesarian section. This, however, leaves scars on the womb and the psyche which cause problems later.

The parents usually decide to conceive again, and most mothers become pregnant soon after the termination as a psychological attempt to replace the lost "child".

#### Chorionic villus sampling (C.V.S.)

Chorionic villus sampling is a relatively new test that has been available since 1983. It can be carried out in the first trimester of pregnancy, unlike fetal blood sampling which is carried out in the second trimester. To carry out prenatal diagnosis at earlier stages of a pregnancy would have been the natural course of improvements in this field. This development, however, was given further impetus by the reservations demonstrated by British Pakistanis about terminating a pregnancy in the second trimester. As the

research findings show, the families in this study had far fewer reservations about termination in the first than in the second trimester. Couples who would not have terminated in the second trimester chose to do so in the first.

Chorionic villus sampling is done at about the ninth to the eleventh week from the last menstrual period. A test done at this stage has both emotional and physical advantages for the mother. The laboratory diagnosis is based on D.N.A. analysis and at present, to affirm whether the test can be carried out for a particular family, family studies based on blood tests are required beforehand. Chorionic villus sampling can usually be done if a normal or thalassaemic child is already present in the family. It may still be possible to have the test if, in the absence of a normal or thalassaemic child, blood can be taken from all four grandparents.

As soon as a pregnancy is reported, an appointment is made for the mother to attend for an ultrasound scan to determine the exact stage of the pregnancy. A further appointment will be made based on the gestational age of the fetus.

The test itself takes 10-15 minutes. So that the uterus can be clearly distinguished under the scan the mother is asked if her bladder is full. If not, she will be asked to drink some water. The proceedings are watched on the ultrasound throughout. Chorionic villus sampling can be done in two

ways. The first method is transcervically using a Portex catheter, a very simple plastic catheter with a metal introducer which is inserted through the vagina and uterine cervix (fig. 1.11).

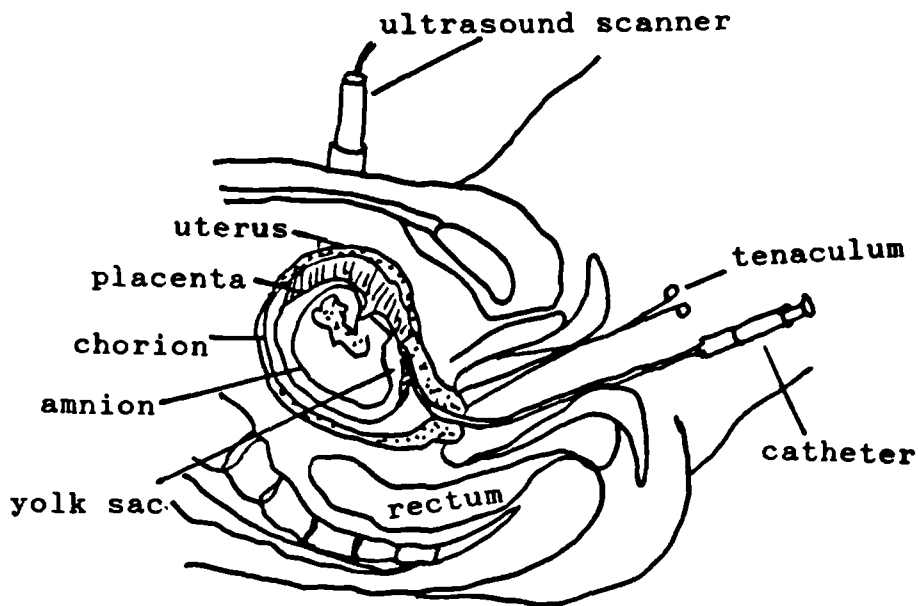


Fig. 1.11: Transcervical chorionic villus sampling.

The tube is so thin that most women hardly feel it. When it is seen on the ultrasound that the tube is in the right position, a syringe is fixed onto the end of the tube to take a small sample of tissue from the placenta. The tissue sample is examined under the microscope to see if it is from the placenta. If it is not, the procedure is repeated immediately, repositioning the catheter. This rarely has to be done more than twice. Alternatively, the procedure is carried out transabdominally (fig. 1.12).

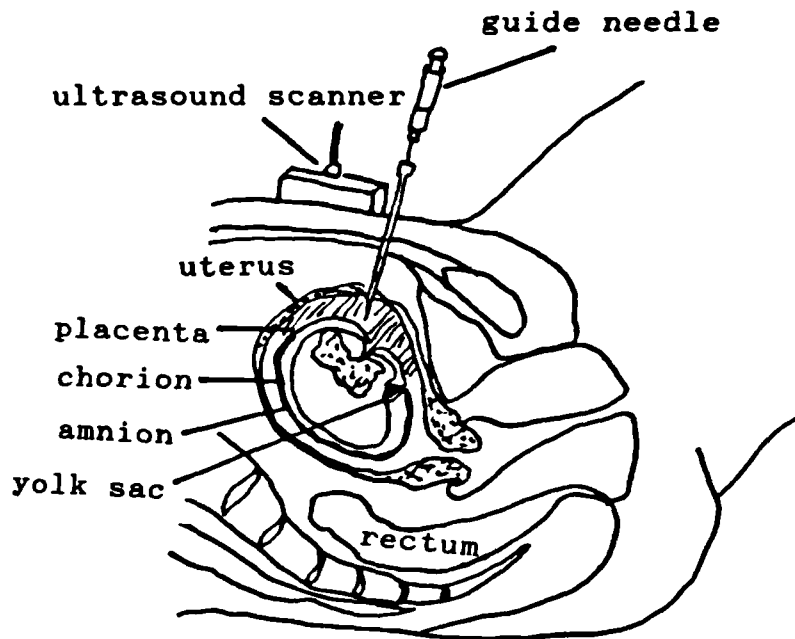


Fig. 1.12: Transabdominal chorionic villus sampling.

Using a local anaesthetic a needle is inserted into the abdomen and placental tissue removed by suction using a syringe. The procedure is observed on ultrasound and repeated if necessary.

Samples obtained are sent to the National Haemoglobinopathy Reference Laboratory in Oxford for D.N.A. analysis. The results take two weeks to obtain, i.e., are usually available before thirteen weeks gestation. Termination at this early stage of pregnancy (less than fourteen weeks) is quick and painless. The mother goes into hospital one evening, and is given a general anaesthetic the next morning. The womb is then emptied through the cervix and the mother is allowed to go home the following day. There is about a 1% chance of making a mistake in this test and the risk of miscarriage is



thought to be 1-2%. This type of termination rarely causes complications in future pregnancies.

Until 1985 prenatal diagnosis was available only in London. Prior to the availability of C.V.S. locally, couples in this study travelled to University College Hospital in London for the test. This entailed the expense of travel and overnight accomodation for the spouse, making arrangements for the care of other children, and loss of privacy because explanations had to be offered to relatives looking after their children during their absence. For the unemployed, applications had to be made for travel warrants.

#### World distribution of thalassaemia

The thalassaemias are found with a high frequency in a vast area extending through the Mediterranean, the Middle East, the Indian Subcontinent, South-East Asia including Southern China, Laos, Thailand, Malaysia and many of the islands (map 1.1).

The estimated global numbers of heterozygotes and global annual births for the major haemoglobinopathies are shown in table 1.2 and 1.3 respectively.

#### Explanation for prevalence of thalassaemia trait

The world distribution of haemoglobinopathies coincides with that of *Falciparum malaria* (map 1.2), and it is thought that



Region	Population in millions Population en millions d'habitants	% of the population heterozygous for: % de la population hétérozygote pour :						Total % of population Pourcentage total de la population	Number of heterozygotes (millions) Nombre d'hétérozygotes (en millions)	
		Sickle-cell traits HbS and C Drépanocytose HbS et C	$\beta$ thalassaemia $\beta$ thal	$\beta$ thalassaemia traits HbE	$\alpha^+$ thalassaemia trait	$\alpha^+$ thalassaemia				
Africa — Afrique										
North — Nord	116	0.3-2.3	1-4	...	...	...	4.8	5.53		
North-East — Nord-Est	38	0	0-1	...	...	...	—	0.33		
East — Est	82	1-40	—	...	...	...	11.2	9.15		
South — Sud	34	—	—	...	...	...	—	—		
Islands — Iles	11	1.5-20	...	...	...	...	8.9	1.0		
Sub-Saharan — Sub saharienne	212	2-40	0-4	...	...	...	23.3	49.4		
Total Africa — Total Afrique	493	0-40	0-4	...	...	...	13.3	65.4		
Americas — Amériques										
North — Nord	257	1.6	—	—	—	—	1.2	3.0		
Central — Centre	97	0.6	—	...	...	...	0.6	0.6		
South — Sud	252	0.5	—	...	...	...	3.6	9.1		
Caribbean — Caraïbes	31	3-12	1-2	—	—	—	9.7	3.0		
Total Americas — Total Amériques	637	0-12	1-2	—	—	—	2.5	15.7		
Asia — Asie										
North-West — Nord Ouest	161	0-20	1-16	—	—	—	3.8	6.1		
South — Sud	924	0-40	1-13	0-10	...	...	4.2	39.0		
North-East — Nord-Est	915	...	...	...	...	...	...	—		
South-East — Sud-Est	663	...	1-10	0-40	(1.9)	...	11.6	76.6		
Total Asia — Total Asie	2 663	0-40	0-16	0-40	0-4	0-4	4.6	121.7		
Europe										
North-West — Nord-Ouest	237	—	—	—	—	—	0.3	0.72		
North-East — Nord-Est	79	...	...	...	...	...	...	...		
South — Sud	172	0-25	1-15	...	—	—	2.4	4.13		
USSR — URSS	270	—	0-15	...	...	...	1.7	4.65		
Total Europe — Total Europe	758	0-25	0-15	—	—	—	1.3	9.5		
Oceania	28	—	—	—	—	—	1.1	0.3		
Global total — Total mondial	4 579						4.6	212.6		

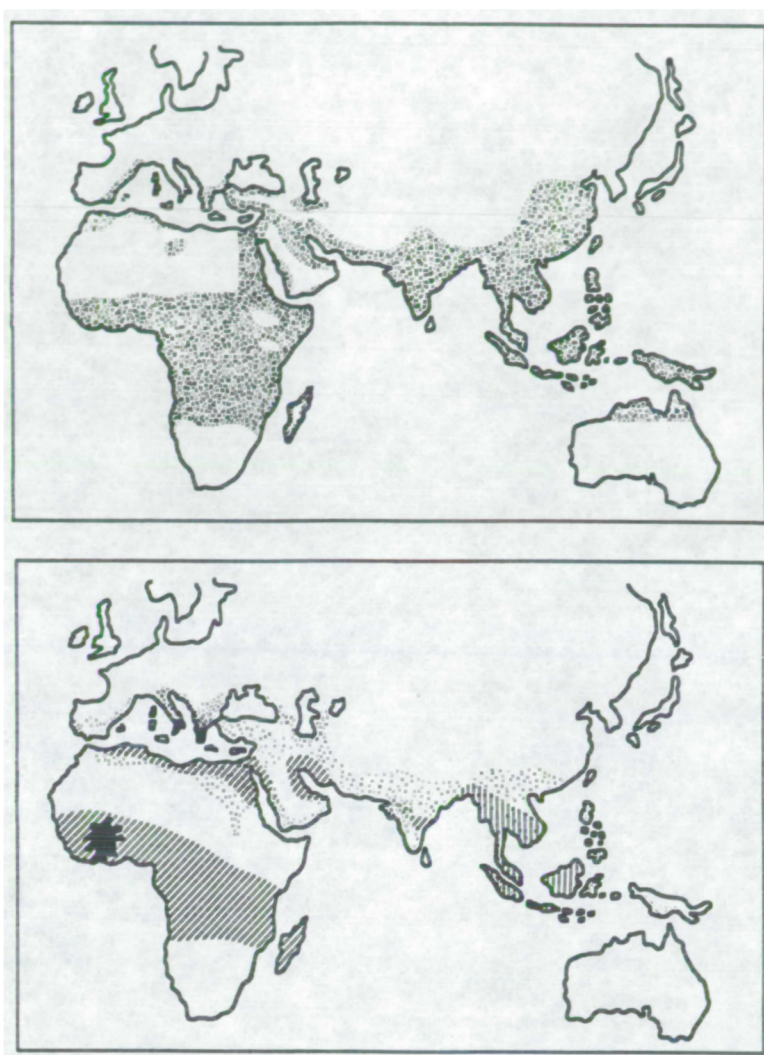
Table 1.2: Global numbers of carriers of haemoglobinopathies

Source: World Health Statistics Quarterly (1988), p.214.

Region - Région	Affected births per 1 000 Naissance d'enfants atteints pour 1 000	Thousands of annual births of homozygotes with Milliers de naissances par an d'homozygotes porteurs de :						Total (000)
		SS	SC	S/β thal	β thal	HbE/thal	α <sup>+</sup> thalassaemia α <sup>+</sup> HbH	
Africa - Afrique								
North - Nord	1.17	[16]	1.17	30	...	...	...	5.77
North East - Nord Est	0.02	...	...	0.04	...	...	...	0.04
East - Est	3.5	13.63	...	...	...	...	...	13.63
South - Sud	...	...	...	...	...	...	...	...
Islands - Iles	2.3	1.05	...	0.01	...	...	...	1.06
Sub Saharan - Sub-saharienne	14.0	110.4	31.6	...	...	...	...	142.0
Total Africa - Total Afrique	9.2	126.7	31.6	>1.2	>3.1	...	...	162.6
Americas - Amériques								
North - Nord	0.25	0.57	0.26	0.13	0.09	...	...	1.05
Central - Centre	0.04	...	...	...	...	...	...	...
South - Sud	0.51	2.28	1.04	0.5	0.4	...	...	4.22
Caribbean - Caraïbes	2.9	1.47	0.67	0.32	...	...	...	2.48
Total Americas - Total Amériques	0.47	4.3	2.0	0.95	0.5	...	...	7.9
Asia - Asie								
North-West - Nord-Ouest	1.28	4.0	...	1.43	3.1	...	...	8.52
South - Sud	0.94	14.81	...	...	14.81	1.84	...	31.46
North-East - Nord-Est	...	...	...	...	...	...	...	...
South East - Sud Est	1.23	...	...	...	7.06	15.3	7.23-16.5	46.1
Total Asia - Total Asie	1.15	18.8	...	1.43	25.0	17.1	23.73	86.1
Europe								
North West - Nord Ouest	0.13	0.28	...	...	0.12	...	...	0.40
North East - Nord-Est	...	...	...	...	...	...	...	...
South - Sud	0.28	...	...	...	0.9	...	...	0.9
USSR - URSS	0.08	...	...	...	0.87	...	...	0.9
Total Europe	0.16	0.3	...	...	1.89	...	...	2.2
Oceania - Océanie								
	0.21	...	...	...	0.1	...	...	0.1
Global total - Total mondial	2.1	150.1	33.6	3.6	30.6	>17.1	23.7	258.7

**Table 1.3: Global annual births of infants with major haemoglobinopathies**

Source: World Health Statistics Quarterly (1988), p.216.



Map 1.2<sup>3</sup>: Distribution of Falciparum malaria (above) compared with the original distribution of the haemoglobinopathies.

Thalassaemia  Sickle cell  HbE  HbC 

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<sup>3</sup>Source: Lehmann and Huntsmann (1974). In Modell and Berdoukas (1984), p.5.

heterozygotes are resistant to this particularly lethal strain.

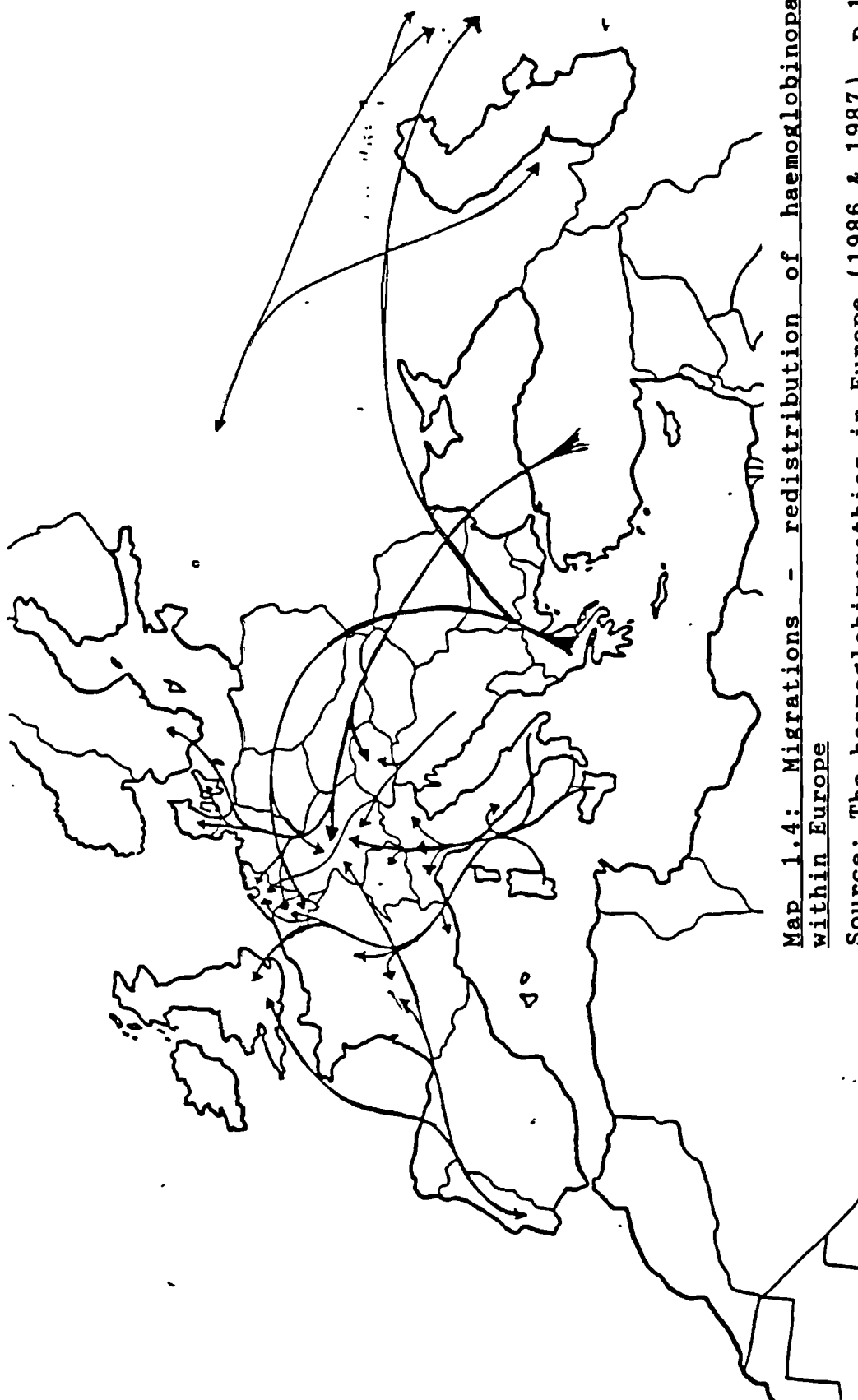
In endemic areas 2-10% of young children used to die from malaria. Those who survive have developed immunity. A characteristic such as thalassaemia trait that protects young children from such a high risk, obviously balances the statistical disadvantage that some children in the population will die from thalassaemia major. In fact, the incidence of the trait in a population is thought to reflect the (previous) mortality from malaria.

Incidence of thalassaemia trait among populations of diverse origins in Britain

Thalassaemia is extremely uncommon among the native British. About one in one thousand individuals is a heterozygote (Cook and Lehman 1973, Knox McAulay et al, 1973) and the expected birth rate of homozygotes is about one in four million, i.e., one per five years. More recently, several haemoglobinopathies, including thalassaemia have been brought to Britain by movements of populations (Maps 1.3 and 1.4).

The approximate incidence of haemoglobinopathies in Britain by ethnic group is shown in table 1.4. Seventeen percent of people of Cypriot origin carry  $\beta$  thalassaemia trait, probably the highest incidence in the world. Greek and Turkish Cypriots are equally affected (Banton 1951a, Modell et al,





Map 1.4: Migrations - redistribution of haemoglobinopathies within Europe

Source: The haemoglobinopathies in Europe (1986 & 1987), p.12.



Ethnic group	Size of pop(approx)	Incidence of		No. homozygote births/yr
		heterozygotes %	homozygotes /1000births	
Cypriot	150,000	17	7.2	22
ASIANS				
Pakistanis		6.5	1-2	6-12
Sikhs	636,000	3.5	0.3	
Bengalis				
$\beta$ thal		3.7	0.3	2
$\beta^E$		3.9		
HbE thal			0.35	
East African Asians				
(Gujaratis+ Sindis)	120,000	10	2.5	6-10
Chinese	70,000	4	0.4	0.6
Malta+				
Gozo	48,000	3.5	0.3	0.3
Italians	164,700	2.5	0.2	0.5
West Indians				
$\beta$ thal		1.5	0.1	0.4
$\beta^S$	604,000	8	1.6	12
HbS thal			0.3	2
Vietnamese	15,000	?	?	?
British	50x10 <sup>6</sup>	0.1	0.00025	0.25

**Table:1.4: Approximate incidence of haemoglobinopathies in Britain by ethnic origin**

Source: Modell and Berdoukas (1984), p.77.

1972). The incidence of thalassaemia trait among the heterogenous population of Asian origin varies markedly from group to group. For example, 14 per cent of some castes of Gujerati origin, about 3 per cent of Sikhs and 3.5 per cent of people of Bengali origin ( Sharma et al 1971, Sidoo et al 1956, Sukumaran and Masters, 1974) carry  $\beta$  thalassaemia trait. 6.5 per cent of people of Pakistani origin are carriers of thalassaemia trait (Dr Hunt, personal communication).

Thalassaemia major births in Britain from 1945 to 1979 are shown in table 1.5.

Originally the majority of thalassaemia births in the UK were to parents of Cypriot origin, but the birth rate of thalassaemic children in this population has declined since 1977, mainly as a result of the introduction of prenatal diagnosis. Affected births have declined particularly in

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<sup>3</sup> Source: Modell and Berdoukas (1984), p.78.

London, as it is here that the majority of people of Cypriot origin live, and preventative methods for thalassaemia were first established. Since the 1960s the populations of Asian origin, particularly British Pakistanis, have contributed an increasing proportion of the thalassaemic births. These have been mainly in areas outside London where people of Asian origin live and preventative services are least available. The place of birth of 364 patients with thalassaemia major are shown in map 1.5. Presently, there are a total of about 300 living thalassaemia major sufferers in Britain.

### THE FAMILIES IN THE STUDY

The families and children with thalassaemia contacted during the course of the research are shown diagrammatically in fig. 1.13. There were thirty one families with a total of forty five thalassaemic children. Forty two of these suffered from thalassaemia major and three had thalassaemia intermedia.

Eleven families with thirteen thalassaemic children were studied intensively: all had thalassaemia major except one who had thalassaemia intermedia. Another three families with a total of five thalassaemia major sufferers and two thalassaemia intermedia sufferers were partially studied. Eight families were met between one to five times, this usually being in a family association meeting. Another eight were met once at home or at a meeting organised by a health professional. In addition I knew of two families to be at-risk for having children with thalassaemia major, and was able to visit one but not the other due to the constraints of time.

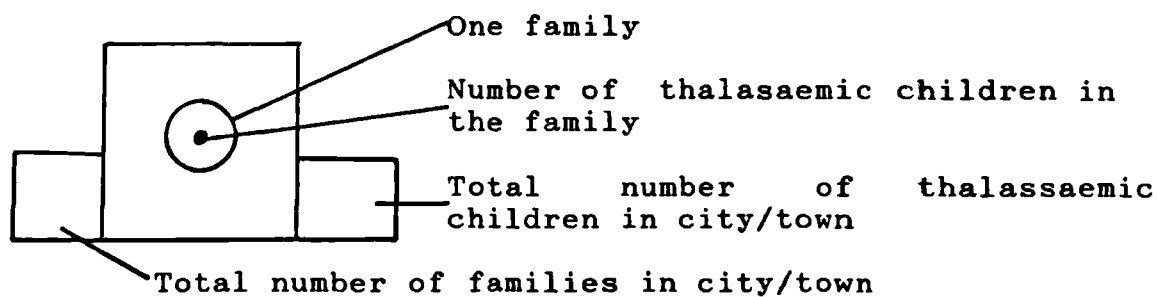
All the above families are Muslims of Pakistani origin. However, I also came into contact with two Sikh couples of Indian origin. One had been prospectively detected as an at-risk couple and the other had a thalassaemic child. Both families were met only on one occasion.

Key to fig. 1.3

- Intensively studied
- Partially studied
- ◐ Met 1-5 times
- △ Met once
- ◇ Contact through telephone conversation

miles Distance from city Z.

----- Families moving residence during course of research



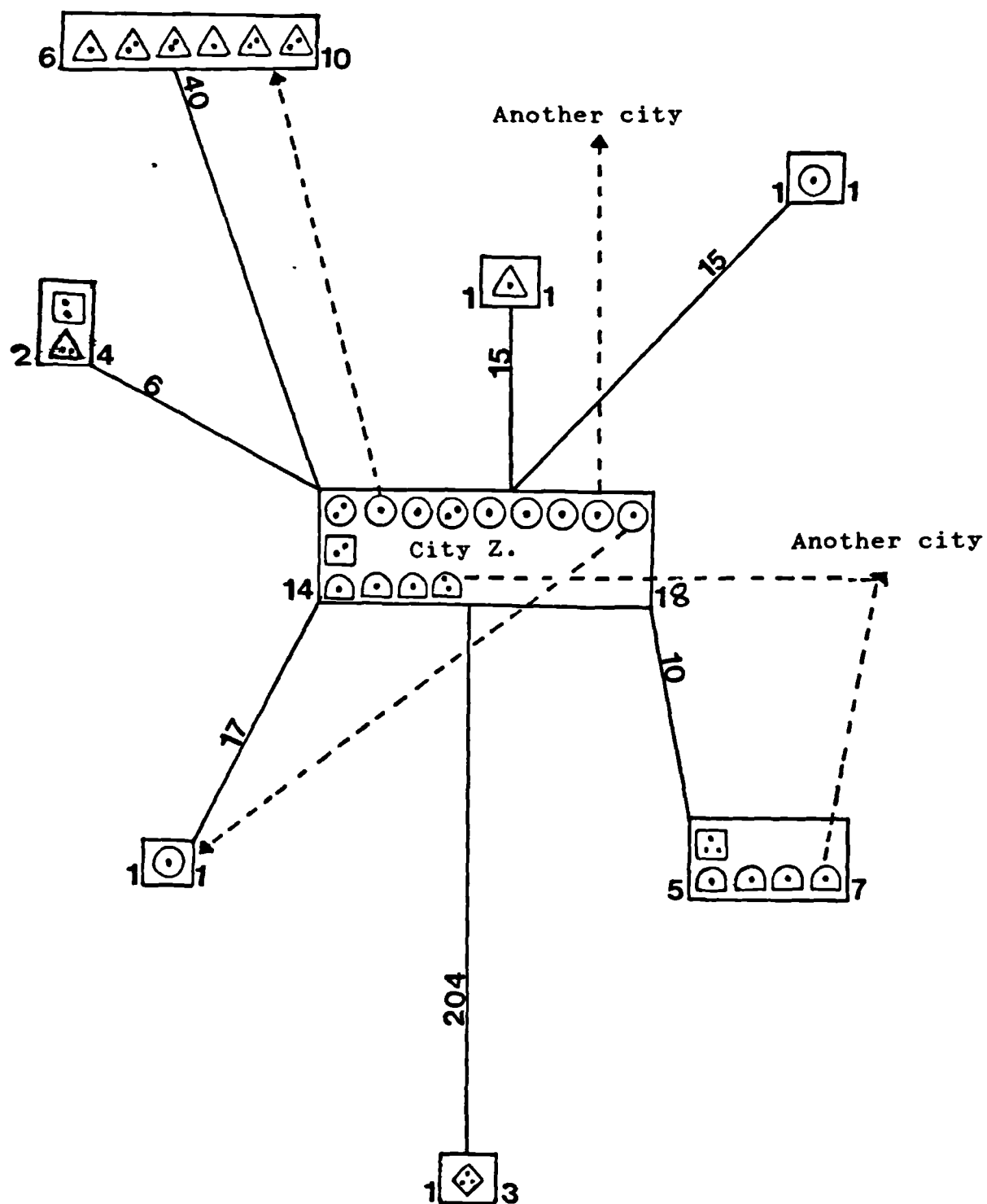


Fig. 1.13: The families in the study

## INTRODUCTION TO THE ELEVEN INTENSIVELY STUDIED FAMILIES IN THE STUDY

The following is a brief introduction to the eleven intensively studied families in this study. Detailed family studies are presented in a later chapter (pp. 173-241). This introduction is meant to serve as an initial contact with the families in the study before proceeding to discuss the wider background of British Pakistanis.

At the beginning of the study in 1981, six of the eleven mothers were in their twenties and five in their early to mid-thirties. Five of the fathers were in their twenties, three in their thirties and two in their forties. The husbands were generally older than their wives and in a few cases of the same age.

All eleven couples were born in Pakistan. Five couples were of urban origin (from the Punjab province); and six of rural origin, two of whom were from the Punjab and four from the Mirpur District of Azad Kashmir. This small sample is not representative of the total population of Pakistani origin in Britain in which 60-70% have their origins in the rural Mirpur District area (Saifullah-Khan, 1977). However, of the total number of families met it can be said that most were from the Mirpur District area.

Three of the women had arrived in England in the 1960s; one came at the age of seven with her mother and siblings to join

her father, the second came at the age of fifteen, again with her mother and siblings and the third came as a newly married woman to join her husband. Six women came in the 1970s, virtually all came to join their husbands, who were already living and working here and had returned to Pakistan to marry. These families are representative of the groupings of women and children who came to Britain in the 1960s and 1970s. Two women arrived in the 1980s; one as a newly married woman joining her husband and the other with her children to join her professional husband who had been posted in England for a few years. All the women are housewives. Only one had been in employment prior to marriage.

Eight of the eleven husbands came to Britain in the 1960s; five as dependants with their father or both parents, one with his newly married wife, one as a married man (his wife remaining in Pakistan) and one as a single man in his late teens. Two men came in the 1970s, both as fiances of British Pakistani women, to marry and settle in Britain. One man arrived in the 1980s, a professional posted in England for a few years. The groupings and arrival dates of the men also represent the general trend of male migration from Pakistan.

Only two of the women were fluent in English; one had been through the U.K. education system from the age of five to fifteen years, the other had spent one year at college learning English prior to taking up employment after her



arrival in England. Both are bilingual. Six of the women had received between six and ten years of schooling in Pakistan<sup>1</sup> and all six were literate in their national language, Urdu. Fluency in English among these six women varied: none were confident about understanding or speaking it, although some were attending English classes. Three of the women had received no schooling, were not literate in Urdu, and spoke only their mother tongue, Punjabi. One of these women, though not literate in Urdu was literate in Punjabi, a language usually reserved for poetry and some religious writings, but generally not used for communication in writing. All of the women were able to read Arabic, having received instruction on the recitation of the the Muslim scripture, the Qur'an.

None of the men were illiterate. Three were graduates, all from urban areas. Of the graduates, one had studied theology for a further seven years in Pakistan after graduating and another had retrained in engineering in England after being

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<sup>1</sup> Primary schooling in Pakistan begins at the age of five. The first public examinations are taken ten years later for the Certificate of Matriculation. The intermediary (F.A./F.Sc.) are taken two years later and this can progress to graduation with a B.A. or B.Sc. with a further two years study. The graduate courses were previously only offered at universities in the major cities, but are now available in colleges of further education, in the smaller cities. Postgraduate courses are available at universities or polytechnical colleges. Educational qualifications from Pakistan are not officially recognised in Britain, though unofficially, a B.A./B.Sc. degree from Pakistan is viewed to be of the same educational standard as G.C.S.E. "A" levels.

made redundant. Both were fluent in English, Urdu and Punjabi and literate in Urdu and English. The other graduate worked in a factory prior to being made redundant and has a working knowledge of English. Two of the men had studied in England for a few years until the age of fifteen and were fluent in English but one was not literate in Urdu. One had matriculated in Pakistan and had a teaching qualification. Five had had between five and eight years of schooling. All five of these men were literate in Urdu though not confident when speaking English. All the men were able to read Arabic for the recitation of the Qur'an.

All the men, except the theologian, had found only unskilled or semi-skilled work. The theologian was employed within the British Pakistani community. The study's time-span (1981-1986) was during a period of economic recession: in 1981 one man was unemployed, two had just been made redundant and four were made redundant during the study period. Only four men were in full time employment throughout the course of the study. Prior to their unemployment most of the men had worked either in the textile mills, in a foundry or in public transport. In general it was felt that the prospects for finding similar work were bleak, and the tendency seemed to be to look for work within the community or to pool resources with kin or friends and consider self-employment.

All eleven families live in terraced houses in the inner city. At the beginning of the study one family lived in a suburban area as a nuclear family, but since moved to an inner city area and now live as an extended family. Nine out of the eleven intensively studied families live as nuclear families. The other two families alternated between living as a nuclear and extended family as family circumstances and finances dictated.

All the families live in areas with a concentration of other families of Pakistani origin, with access to shops and amenities catering for their specific ethnic tastes, for example, grocers, halal butchers, fabric shops, Pakistani banks and travel agents. Kin and friends often live in the same or neighbouring areas. In city Z., as in most industrial cities, the inner city areas are made up of rows of large and small terraced houses. These houses were built either before or just after the turn of the century; the larger ones for large victorian families and the medium-sized and smaller "back to back" houses were built to accommodate the influx of workers from the rural parts of England. These inner city areas have always provided cheaper housing for immigrants until they prosper and move out, and a new wave of immigrants take their place. In this study there were families living in a large house with only two children and in a small house with a much larger number of children; the house size did not necessarily correlate with the needs of the family but was

rather determined by what the parents were in a position to afford.

None of the families owned a car. They all had to depend on public transport to attend hospital appointments and meetings unless a relative or friend was able to offer them a lift. Six of the families did not have a telephone at the beginning of the study; one family acquired one when they moved house. The absence of the telephone meant making a trip to a public phonebox to contact the hospital, doctor or me and repeated visits if the first trip was unsuccessful. None of the homes were centrally-heated. One family had not been able to afford a refrigerator, and eventually acquired one from the Social Services' department because they needed it to store the iron-chelating drug, Desferal. It is largely in the context of economic insecurity, and bad and overcrowded housing that these families are coping with chronic disease.

### MIGRATION TO BRITAIN

This chapter describes the context of Pakistani migration to Britain, giving an indication of the social and political climate at the time of their entry. It also points to the trend in Britain towards the development of a multicultural society, changing the face of its population and precipitating a need for adaptations in service provision.

Migration of labour between countries, dictated by socio-economic and political factors, has become a feature of the modern world with implications at various levels. It was with the growth of capitalism and the Industrial Revolution that migration of labour, primarily for economic reasons, arose from the seventeenth century to the present. The process is a continuing one and has created plural societies in that nations now consist of different ethnic and racial groups (Anwar, 1985, p.1).

Britain was one of the first countries to have large scale migration of labour in the eighteenth and nineteenth century. The surplus of unemployed workers from the countryside was soon absorbed by the Industrial Revolution. British employers then turned to Ireland to fulfill their need for labour (Thomas, 1968, pp.469-85). In 1851 Irish migrants made up 2.9 per cent of the population of England and Wales (Jackson, 1963, p.14). Twelve thousand Jewish people came to Britain between 1875 and 1914, escaping persecution in Eastern Europe.

Approximately 460,000 migrants entered Britain between 1946 and 1951, the largest group being the 115,000 Poles who came under the Polish Resettlement Scheme. A smaller number of prisoners of war, mainly Italians, Germans and Ukrainians were allowed to settle in Britain (Isaac, 1954). Ninety thousand European Voluntary Workers (E.V.W.s) were also recruited from refugee camps and given three year contracts for a specific job.

Migration from the Commonwealth countries, which includes Pakistan, began as a trickle after the Second World War, responding to the demand for labour in Britain. Citizens of the Commonwealth countries had free entry into Britain under the Commonwealth rules. Until the early 1960s all the migrants from the Commonwealth were economically active, the first wave of migration being mainly women from the West Indies. This was followed by people from India and subsequently by Pakistanis who began to migrate in sizeable numbers in the mid-1950s. Migrants from India and Pakistan were predominantly men. The government of Britain had actively encouraged this immigration, setting up recruitment agencies in the migrants' home countries. The jobs available were in the declining industries with poor conditions.

The influx of cheap migrant labour slowed down the final closure of certain declining industries and pre-empted the need to replace obsolete labour-intensive machinery.

Historically, all foreigners have experienced hostility from the indigenous British population and this often meant exclusion from or segregation within workplaces. "No Irish" signs were subsequently replaced by "No Jews" followed by "No Coloureds". Discrimination in housing and employment were a regular feature of life until outlawed by legislation. Mass demonstrations against immigration resulted in Britain's first restrictive legislation on immigration, the Alien's Act of 1914. Similar pressure in subsequent decades, focussing particularly on the presence of a non-white population in Britain, culminated in the Commonwealth Immigrants Act of 1962, designed to control the influx of non-European immigration (Anwar, 1985, p.4).

Immigration from Pakistan, Bangladesh, India and the West Indies between 1955-68 is shown in table 1.5.

Indians	200,130
Pakistanis (inc. Bangladeshis)	145,960
Jamaicans	191,330
Rest of Caribbean	132,220
Total	669,640

Table 1.6<sup>1</sup>: Net immigration from India, Pakistan, and the West Indies between 1955 and 1968.

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<sup>1</sup> Source: Nicholas Deakin, 1972, p 50. The number of people arriving for settlement from all the New Commonwealth countries in 1969-77 was 318,512 (out of these 259,646 came as dependants and only 58,875 were male workers). This means a drastic decline in the number of immigrants coming as workers from the New Commonwealth countries.

The voucher system, under the Act, still allowed labour to be brought in for the declining industries: a voucher could only be issued by an employer for a vacant job which could not be filled by someone from the local population. As the jobs already held by migrants were those undesired by the indigenous British, migrants began to arrange jobs and vouchers for their relatives and friends. Dependants of those already in employment were allowed entry without vouchers. Many dependants entered the country in an attempt to "beat the ban" as more restrictive legislation was anticipated. Between July 1962 and December 1968 257,220 dependants from the Commonwealth were admitted to Britain compared with 77,966 with vouchers (Anwar, 1985). In 1962 the voucher system was halted with the passing of the Commonwealth Immigrants Act of that year. Since then it has been extremely difficult for males of non-European origin to gain entry to Britain.

"Pakistani migration co-incided with hardening public attitudes, increasingly restrictive immigration controls and severe economic and employment difficulties throughout the country (Saifullah Khan, 1977, p.79)."

At the moment migration to Britain consists only of dependants whose fathers, husbands or sons live here.



### Pakistani migration to Britain

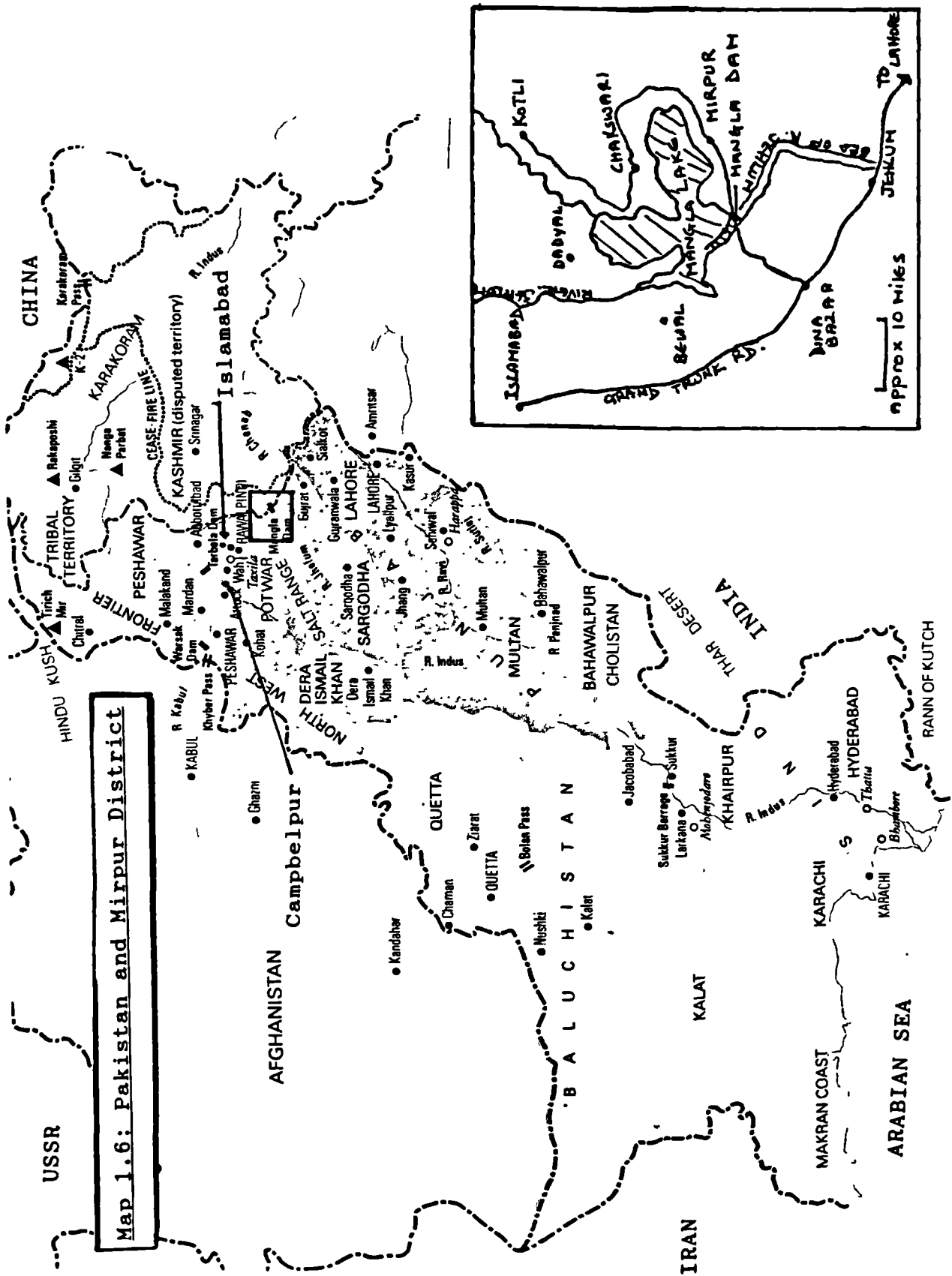
British Pakistanis are not a homogenous population. They share a common nationality and religion but are differentiated on ethnic/regional and class lines. In Britain there are five main sections of the population:

1. People from Mirpur District in Azad Kashmir<sup>2</sup>  
(see map 1.6).
2. People from the Campbellpur District of Punjab Province.
3. Punjabis from other districts of the Punjab (e.g., Rawalpindi, Jhelum, Gujrat, Lyallpur).
4. People from the North-West Frontier Province (N.W.F.P.).
5. The urban educated section from the cities in all the regions.

Sixty to seventy percent of British Pakistanis originate from the Mirpur District of Azad Kashmir, the heaviest migration being from Tehsil Dadyal, a sub-district of Mirpur District (see map 1, inset). The next largest group are from Campbellpur District. Both Mirpur and Campbellpur Districts are essentially Punjabi in culture. The third largest group are from the other Districts of Punjab. In total then the vast majority of British Pakistanis are culturally of Punjabi origin with smaller numbers from the North-West Frontier Province and the cities (Ballard, 1986, Saifullah-Khan, 1977).

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<sup>2</sup> Azad Kashmir is the Pakistani held territory of the previous state of Jammu and Kashmir. The other part of the old state is held by India and the whole region, separated by a ceasefire line, remains disputed territory.



Some of the reasons for coming to Britain of a sample of 103 British Pakistanis interviewed by Anwar (1985) are listed in table 1.6. From the literature this reflects the reasons of the migrant population as a whole. The main motive for migration was economic.

The earliest Pakistani migrants were men, migrating in the 1930s from a scatter of villages in what is now Mirpur District. These ex-seamen were the principle pioneers of the present settlement of British-Pakistanis in Britain and took jobs as stokers in the British Merchant ships at the turn of the century. It is as a result of their migration that at least three quarters of British Pakistanis have their origin in an area twenty miles by thirty miles, lying mostly in Azad Kashmir with a particular focus on Mirpur District. Half or more of the population from these villages, lying closest to where the seamen were recruited, now lives in Britain (Ballard, 1986). Many Punjabis had also been recruited into the British Army in India as Punjabis were thought to be a 'particularly martial and loyal race'.

When the seamen came ashore most earned a living as pedlars, hardly any industrial jobs being available. It was after the Second World War that the numbers of pioneers grew rapidly. Britain's industries were chronically short of labour and former pedlars took on industrial jobs. During the late 1940s and 50s increasing numbers of seamen left their ships to take

industrial jobs on shore. Seamen whose ships had been torpedoed beneath them were drafted into munitions factories in Yorkshire and the West Midlands (Dahya, 1973). After the war some returned to Pakistan with accumulated savings whilst others remained to carry on working. Stories circulated about the economic prospects in Britain and the seamen began to call over their kinsmen and fellow villagers to join them, thereby setting in motion chain migration.

<u>Reasons</u>	<u>No.</u>
Economic	81
Education	10
To join parents	5
Visit	5
Political	1
Other (Air Force deserter)	1
Total	103

Table 1.7<sup>3</sup>: Main reasons for coming to Britain of 103 British Pakistani men.

Except for the earliest settlers, whether emigration was even a possibility for a family member was determined by whether there was already a member of the family or village working in Britain. Rarely did anyone venture out without a prior contact with someone who would facilitate the passage into life in Britain. The decision to migrate was not taken by an

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<sup>3</sup>Source: Anwar (1985), p.25.

individual, but by the head of the household and kin who would finance the trip and expected to benefit from the investment. The distance produced by the migration was not expected to diminish the relationship between the migrant and his family in the village. Conversely, bonds of affection and obligation were further increased as a person's passage was facilitated to make a contribution to the combined resources of the household. These responsibilities increased the migrant's respect in the family and his influence in family decisions. Despite the migration the strong kinship networks persisted between the two countries.

Due to the numbers migrating, by the 1960s the institutions of migration such as travel agents and banks had become firmly established in both countries, and a new air route between Islamabad and London had been opened to deal with the traffic (Ballard, 1986, Saifullah-Khan, 1977).

In the 1960s the pattern of migration shifted. Anwar (1985) states that the 1962 Commonwealth Immigration Act had a decisive effect on the pattern of migration and turned a movement of workers who were probably only interested in staying temporarily into a relatively permanent immigration of families. The reuniting of families was also more feasible by this time, as by the 1960s most men were able to afford suitable living conditions for their wives and children. This point is particularly relevant to Pakistani migration, where

up to 1962, virtually all the migrants had been men. The ratio of Pakistani males to females between 1961 and 1971 is shown in table 1.8. The ratio of males to females in 1982 was 58% males to 42% females. For the general population in the same year it was 48% males to 52% females (Anwar, 1982).

In 1962 when the issue of vouchers was halted it became extremely difficult for males of non-European origin to gain entry to Britain, even though jobs were plentiful. The voucher system had reinforced the kinship and friendship bonds and affected patterns of settlement as the earlier migrants had found jobs for their kin and friends in the factories where they themselves worked, increasing the element of patronage.

	<u>Males per 100 females</u>
1961	538
1966	354
1971	295

Table 1.8<sup>4</sup>: Pakistani sex ratio between 1961 and 1971

Between the mid-50s and early 70s the migrating males usually left their wives and children behind in the care of other family members. This was made easier for migrants with consanguineous marriage partners as their wives would be

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<sup>4</sup> Source: Prepared by Anwar, 1985, p 39, from information in tables in G B Lomas, Census, 1971, The Coloured Population of Great Britain (The Runnymede Trust, 1973), p 80.

living with people who were already their kin. Often the wives looked after the elderly parents. In this period the men behaved as "international commuters" making repeated and lengthy visits to their villages, during which time they assured themselves of their family's welfare, spent time with their wife and children and reconstructed their house. Often a wife would be pregnant before her husband left again for Britain. As employment levels were high it was not difficult to slot back into the labour market on return. The economic decline of the late 1970s, particularly affecting the textile and heavy engineering industries where most men of Pakistani origin were employed, created high unemployment among the workers from those industries. Families once settled in Britain tend to make persistent but less frequent visits to Pakistan.

Migration from Pakistan now comprises families who are still to be reunited and the spouses and children of British adults of Pakistani origin who returned to Pakistan to marry, plus families who have lived in Britain but subsequently returned to Pakistan, and now wish to settle in Britain once again. Some earlier settlers now of pensionable age have returned to Pakistan.

The passage to Britain may initially have introduced a large physical gap between the migrants and the remainder of their kinship networks in Pakistan and made them lose their economic

niche there, but it did not necessarily lead to any great disjunction in the structure of those networks themselves. Those who moved overseas continued to see themselves as family members, for their initial object in migrating was not to settle permanently overseas, but to add to their families' corporate resources by taking temporary employment. Chain migration ensured not only that most migrants were soon surrounded by a wide range of kinsmen and neighbours, but also that they were followed by several other members of their immediate corporate families (Ballard 1988).

The bulk of Pakistani migration, mediated through the kinship system, has been highly selective and localised. It has been the transference of pockets of population from specific areas of Pakistan to specific parts of Britain. Sponsorship and patronage are crucial elements in the understanding of this process (Anwar, 1985). Pakistani migration has not been a single time-limited episode. Instead kinship networks, changing patterns of material opportunity and modern travel facilities have led to the establishment of permanent links between the British-based and the Pakistan-based components of a single community. Constant travel and communication back and forth are indicators of this trend. As communications have become cheaper, so the traffic as well as the bond between Britain and parts of Pakistan, particularly Mirpur, is tending to be consolidated. British Pakistanis now operate, as they always have, within a social arena which encompasses



both Pakistan and Britain; and they move around within this arena as events and circumstances dictate.

This feature of maintaining and utilising kinship-friendship networks, also characteristic of some other migrating populations, figures very strongly in the cultural make-up of British Pakistanis even now. The understanding of this feature is essential in an analysis of the British Pakistani situation and, as will be discussed later, has important implications for the delivery of a genetic service to this population.

PART TWO: SOCIAL BACKGROUND

It is important to set out the societal background of the families in this study, both in Pakistan and in Britain. In order to understand fully an individual's or group's situation in an adopted country a qualitative study must include the socio-economic, political and cultural forces in the emigrants' country of origin, as well as the forces in the wider community in which they have settled. The conditions giving rise to decisions both at a societal and individual level are a response to these forces at various levels. As a true picture can only emerge if all these factors are taken into consideration, the next part of this thesis therefore describes the salient features of the background of British Pakistanis in Pakistan and Britain, which are pertinent to understanding the experiences of families to be presented later.

## PAKISTAN

The area which now constitutes the state of Pakistan was part of India before India's independence from British colonial rule in 1947. Pakistan was created due to pressure from the muslims in India for an independent muslim state and in 1947 comprised two geographically separate parts - East and West Pakistan, with 1,000 miles of Indian territory in between. The civil war in 1971 between the two wings of Pakistan resulted in East Pakistan declaring itself the independent state of Bangladesh. Except for a common religion, Islam, the two populations differed considerably from one another.

Pakistan now consists of four provinces; Sind, Baluchistan, the North-West Frontier Province (N.W.F.P.) and the Punjab, plus the disputed territory of Azad Kashmir. The capital of Pakistan, Islamabad lies in the Punjab (see map 1.6, p. 95). The Punjab Province was split during the partition with mass migrations taking place from both sides - the Hindus and Sikhs moving to East Punjab in India and the muslims to the West Punjab in Pakistan. Mass migrations also took place from other states, particularly those surrounding the borders. These migrations have also meant the movements of genes with the people.

Azad Kashmir is the Pakistani-held territory of the previous state of Jammu and Kashmir. The other part of the old state is held by India and the whole region, separated by a

ceasefire line, remains disputed territory. Azad Kashmir's history and socio-economic conditions plus its unusual relationship with Pakistan are presented later (pp. 4-5) as background to information on the Mirpur District.

The four provinces of Pakistan each have their own distinctive culture and language. Whilst the national language, Urdu, is used in schools as the medium of teaching and in Government offices, the regional language is usually spoken in the home. Hence Pakistanis are a heterogeneous population, their religion, Islam, and a common national history being the unifying factors. Virtually all Pakistanis are muslims, the vast majority being of the sunni sect<sup>1</sup>. Jewish, Christian, Hindu, Sikh and Zoroastrian people also make up a small minority of the population. The climate of Pakistan is varied, spanning the heat of the deserts in Sind and the colder temperate climate of the Northern mountainous regions.

The areas attributed to Pakistan on the partition of India consisted mainly of undeveloped, agrarian societies. Eighty per cent of Pakistan's population of 100 millions (1987 figures) live in the villages and twenty per cent in the urban areas (Anwar, 1985). A largely ineffective birth control programme has meant a rapidly increasing population estimated at 150 millions by the year 2000 if present trends continue

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<sup>1</sup> Islam has two major religious sects, the sunnis and the shia's. The sunnis form the majority sect in Pakistan and the Islamic world as a whole.

(Hyman et al, 1988).

In the forty two years since its creation Pakistan has undergone politically turbulent times and remains a nation still trying to find its feet both politically and economically. It is extremely rich in natural resources but is currently facing an economic crisis (Ahmed and Amjad 1984). An important feature of its economy are remittances from workers overseas, which account for over half of Pakistan's income in foreign exchange. Approximately ten per cent (2.5 millions) of Pakistan's male adult manpower works overseas, mainly in the oil-producing states of the Middle East. These migrants are of similar origins to those who migrated to Britain in the earlier decades. Since the mid-seventies migration has played an increasingly important role in the national economy (Ballard, 1988). However, despite the income from remittances deposited in the rural banks, for reasons peculiar to Pakistan's political order (see Hyman et al, 1988) infrastructural development in the rural areas, for example, on roads, canals, electrification, education and health care has been minimal. The major cities, though, have better provision. Facilities are concentrated here and city life has a superficial air of prosperity. Imported consumer goods such as cars, televisions, washing machines and air conditioners are freely available for those who have the means to buy.

There is close and frequent contact between the city and the

village in Pakistan, as many of the twenty per cent who live in urban areas are first-generation urban dwellers who may still have relatives or property in their village of origin. As well as international migration, internal (rural-urban) migration is also a long standing feature in Pakistan. Both types of migration have been, and are still, characteristic of every region to varying degrees.

### Azad Kashmir

The state of Jammu and Kashmir, now Azad Kashmir and the disputed territory of Jammu and Kashmir held by India, is bounded by Pakistan, China, Afghanistan and India with a population of 6 millions (see map 1.6, p. 95). The old state was a combination of half a dozen regions all with their own disparate topography, climate and language. These include the Dogri speaking Jammu in the South-East, the Punjabi speaking South-Western region (where Mirpur District lies), the western part which blends with Hazara in Pakistan, Dardistan and Baltistan in the north and Ladakh comprising the eastern part. In the middle flanked by range after range of mountains lies the famous Vale of Kashmir. The scenic beauty of the land with its mountains, lakes and woodland has prompted writers to refer to it as "heaven's garden on earth". In contrast to its tranquil beauty, Jammu and Kashmir's political history has been tempestuous. Long periods of autonomy, assertion and flourishing have alternated with devastating periods of tyranny and exploitation. In the more recent past, during

British rule of the subcontinent, Jammu and Kashmir remained an independant state. In 1931 Jammu and Kashmir saw mass agitation erupt against Hindu Dogra rule which had done little to improve the poor living conditions. The agitators were supported by the muslims of the subcontinent. The resistance movement peaked in 1946-47, coinciding with the independance and partition of the subcontinent. Both India and Pakistan sought control of the state for emotional and religious reasons, particularly as it holds the headwaters of the subcontinent's major rivers. In 1948 the United Nations Commission intervened and fourteen months of battle ended in ceasefire. Since then, the United Nations has maintained a peacekeeping force along the ceasefire line.

Azad Kashmir is independant of Pakistan in theory but in reality it is governed by the Ministry of Azad Kashmir Affairs in Pakistan's capital city, Islamabad. Azad Kashmir also claims sovreignty over Indian-held Kashmir. Two wars between Pakistan and India since 1947 over Kashmir territory have created instability in the region. By contrast with British India where land ownership was reorganised, the education system extended, taxes collected and irrigation schemes established to enable the colonisation of new areas, Jammu and Kashmir remained undeveloped. It was not untill the 1970s that health and education systems were introduced.



Mirpur District

Mirpur District is very picturesque, lying in the foothills, flanked by the Karakorum range of mountains to the North with the more fertile plains of the Punjab Province to the South (see map 1.6 inset, p. 95). The climate of the region varies with very hot summers and much colder weather in the winter months. Azad Kashmiris generally have a robust constitution, their staple diet being unleavened bread, pulses, meat, vegetables and rice.

Overall, Mirpur District is a rural, unirrigated wheat growing area. It is less prosperous than the irrigated southern plains of the Punjab but has always been densely populated. A major development affecting Mirpur District was the construction of the Mangla Dam. In the 1950s, a joint international venture (U.K., Canada, Australia, New Zealand, Germany and the U.S.A.) started to build the world's largest earth dam here. This caused a large movement of population, affecting approximately 200 villages and a small town, now called Old Mirpur, of approximately 12,000 inhabitants, as well a physical structural remoulding of the area. At the beginning of the 1960s the population near the dam itself was evacuated, from 1963 onwards the whole population of the area was shifted in stages, and by 1967 about 100,000 people (18,000 families) had been moved. Families received compensation in cash for their houses and for those with under half an acre, cash for their land. Farmers with larger

plots could exchange land for land in the Punjab. The number of new villages that have appeared around the lake and the growth of the old villages indicates that most villagers chose to remain as near as possible to their old home and thus near relatives unaffected by the dam. The new Mirpur City grew rapidly in size and population and large villages took in many displaced persons. Many totally new villages arose, often housing the majority of one particular village which had been inundated. Some writers have suggested (Deakin, 1970, p.46, Allen, 1971, p.32) that the large number of people from Mirpur in Britain is a direct result of displacement by Mangla Dam, and an arrangement at governmental level to admit them into Britain. Although this was a contributory factor, migration to Britain had begun even before the displacement (Saifullah Khan, 1977, p.67).

Villages in the Punjab vary in size from a few hundred to several thousand inhabitants and their social structure varies considerably from settlements dominated by large landowners to others with a large number of medium to small landowners. The inhabitants can be divided into those with land and without land. Landowners may own from two to forty acres and some small landowners work as tenant farmers or labourers to supplement inadequate income from their land. Those who do not own land include hereditary craftsmen, village artisans, and tenant farmers. A further category of people cutting across these divisions are villagers who arrived during or

after the partition from India.

Due to the land inheritance pattern in Pakistan whereby all the offspring inherit portions of the parental assets (men inherit twice as much as women), the size of agricultural holdings are small. This factor combined with the conventional division of labour between the sexes has meant that many households have long had a surplus of male labour. There is a long tradition for men from such households to supplement their incomes by working on other farms, in the cities or overseas. Those who migrated to Britain belong to the category of small landholders who wished to supplement their incomes and who also had access to some funds for the journey. In the village they would consider it below their station to do labouring jobs, but working as labourers in factories and mills in Britain was not regarded with the same distaste. The migrants were working for European wages and the position was seen as temporary and a means to an end, that is, to increase the wealth and status in the village setting, status in the village being of paramount importance. As Dahya (1973,) states:

"In the case of the landless migration is internal, while in the case of the small landholder, it is external. Further, the migration is in case related to different ends, and the overseas migrants cannot be lumped with the landless in Pakistan nor can they, in objective terms, be bracketed with the industrial urban proletariat in this country (p.50)."

Migrants to Britain from Mirpur District were village-based

and have had little experience of city life even in their homeland. Due to the lack of educational facilities the formal level of education for all but the rich landowners was less than the average of their countrymen.

"Until recently, in the villages basic literacy in Urdu was the most that most young men achieved, and in fact needed, for agricultural work or jobs overseas (Ballard, 1986, p.12)."

Prior to the impact of remittances from large scale migration in the 1950s, villages in Mirpur District were much the same as villages in other parts of the subcontinent: the compact cluster of houses was divided by irregular alleys which opened out into one or two main lanes or streets. Most houses consisted of two or three rooms leading into a walled courtyard with animal shelters and an open-air kitchen. The courtyard would be connected to a village lane by a high door. Much of the daily life of the household took place in the courtyard, and in the summer the villagers slept outside or on the flat rooftops. It is from villages and houses structured in this way that the early migrants came. Their wives, invariably from the same region, would have grown up in the same surroundings and often in the same village. Men's work was outside of the home working on the land. Women's lives centred around the home, caring for the family, keeping the house and domestic animals in order, cutting fodder and helping in the fields during the busy harvest period. As modern labour saving devices were virtually non-existent, water was collected from the wells, clothes

washed in the river and flour ground with a manual mill (Saifullah Khan, 1977, pp.77-78). For British Pakistanis now in their thirties, Mirpur District holds different memories as the standard of living of the families was raised by the remittances from overseas workers.

#### The impact of remittances

The emigration to Britain has divided villagers into those who have close contact with Britain, other North European countries and the Gulf States, and those without such contacts. The emigration has affected the villagers' awareness of new horizons and alternative lifestyles, introducing a new perspective to village life. Money has largely been invested in the building of larger houses on the edge of their villages or in the new Mirpur City which has been an exceptionally beautiful site for houses built by overseas migrants and the large number of banks and travel agents servicing the migration. The displacement of populations by the lake and the movement of some migrant families to the city has produced a population with close connections to the villages and, for most families, migrant members in Britain. The potentially disrupting influence is modified by the value attached to ancestral landholdings and ascribed status which remains strong, though landowners who have not benefited from emigration experienced a threat to their traditional influence and power (Saifullah Khan, 1977).

Following an economic boom, largely due to overseas remittances, almost every family in the area rebuilt their house in steel and concrete. Having paid the cost of their passage the migrant's next step was usually to improve his family's accomodation. With the sudden inflow of funds and heavy demand for construction materials, the bazars expanded rapidly. Dahya (1973), describes the structural changes in the buildings in Mirpur District and villages in other areas from where migration has taken place:

"As one crosses the Mangla Lake from Mirpur, villages of origin appear dotted all over the horizon in different directions and everywhere "pakka"<sup>2</sup> houses stand out as manifest evidence of the material success of the migrant's families. Similarly in the Chach area, in all the villages to the north east, and west of Hazro town, there are pakka houses which speak for the migrant's economic success. Non migrant families live in "kachcha" houses. Situated at the north-east corner of the Chach is Gurghushti, a village with a population of some 15,000 of which nearly 6,000 have settled in Birmingham, with a few in places such as Bradford and Manchester. From a distance the village appears to be a city of skyscrapers (p.255)."

The areas in which these villages lie can be described as capital rich, with large sums deposited in the banks. In the past two decades large new houses have been constructed by the thousand, bazaars have expanded dramatically, and televisions and videos have arrived in the remotest rural areas. But the underlying structure of the national economy remains unchanged. In all the emigrant areas the capital resources needed to build new roads and irrigation systems,

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<sup>2</sup> A "pakka" house denotes a brick-built house whilst a "kachcha" house is one made of mud.

and to finance electrification and agricultural extension programmes and so forth are available, but public investment in these areas has not taken place. Although at the national level agricultural production has increased, in Mirpur and throughout the unirrigated areas, given the absence of infrastructure support from the state, farming has become less productive, despite the potential fertility of the soil. Farming is no longer regarded as a desirable occupation by returned migrants or their families. (Ballard, 1988).

With increased consumer spending power education has become affordable for families in Mirpur District, and so for the first time large numbers of boys have been sent to school and college. As overseas jobs have dried up, families hoped that education would improve their son's opportunities for employment, but the few local jobs as teachers and bank clerks have long since been filled and there is no local industry.

So although most British Pakistanis now have links with and return to villages with good housing conditions and consumer goods, nevertheless educational and health care facilities in these villages still remain inadequate.

## SOCIAL ORGANISATION

### The Household

The primary social unit in Pakistani society in Pakistan and in Britain is the household. Within the household each position:

"Comprises a complex of rights and duties, attitudes, expectations and sentiments which are balanced carefully to ensure the effective functioning of the unit. Roles are precisely defined into an interlocking pattern of mutual interdependence and individual subordination to the group (Saifullah-Khan, 1977, p.61).

The household may be of many types (fig.2.1):

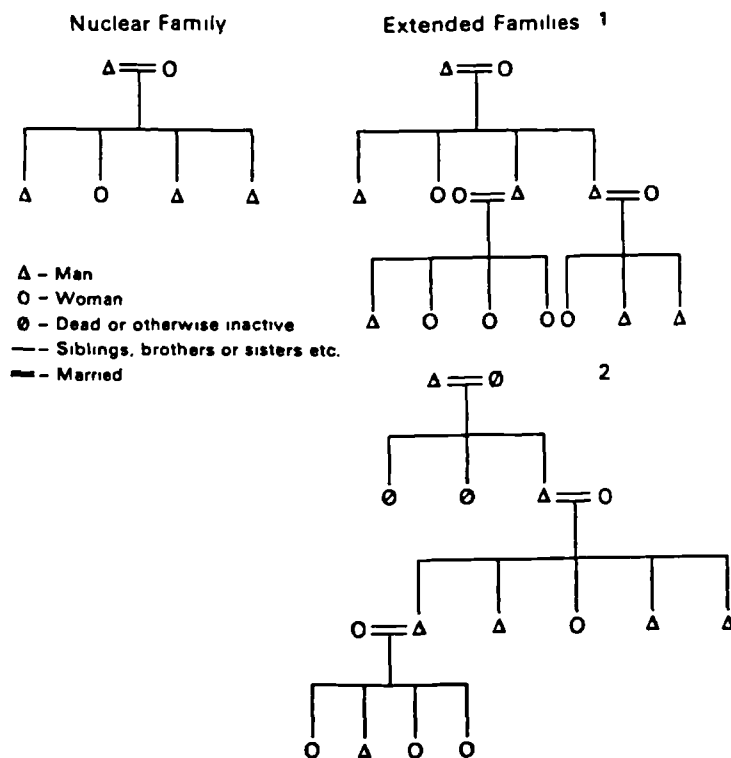


Fig.2.1<sup>1</sup>: Types of household

<sup>1</sup>Source: Anwar, M. (1985), p.53.



Anwar provides a succinct description of the people that make up the varying types of households:

"In the village a family may consist of members of a conjugal family (husband, wife and children) if the married son lives apart from his parents and siblings. It may include all members of a joint family, i.e., the married sons, their wives and children, unmarried sons and daughters and occasionally a widowed daughter. It could be a stem family, the aged parents living with one of their sons and his wife and children, the other grown-up sons having established their own households. Another type is where several brothers live in one compound, with the married brother taking responsibility for their education and arranging their marriages. Married brothers may form separate households when their father dies, but keep their land and business in common (see Mayer, 1960, p.177). The joint family members usually pool their resources and have common property. There could be some absent members of the family who work in a city or live in another country, such as Britain, but have the same status. No doubt, family composition changes over time. It changes according to the natural cycle of any family; some members die, others are born, daughters marry out and daughter-in-laws come in, as it is a patrilocal family structure (Anwar, 1985, p.53-54)."

In Pakistani cities one finds similar patterns of family structure, though modified by the process of urbanisation. Hence there are fewer four generation families and an increased number of nuclear families. However, the three generation family where at least one of the married sons and his family live with the parents in the parental home, with other siblings forming nuclear families, is still the norm for the majority of the urban population. It is rare to find aged parents living alone.

Among British Pakistanis there are fewer extended families than in Pakistan. A national study (Anwar, 1978) found that of 549 muslim (mostly of Pakistani origin) interviewed, 71%

were living as nuclear families and 29 as extended families.

"The households are usually of two generations, compared to the three or more found in Pakistan. Grandparents are less frequently found in Britain, although sometimes they visit their children or come as dependants. When more families have grandchildren born in this country the composition of the household may change.....in some cases married brothers live together and in fact pool their resources as a joint family....another type of Pakistani household exists in Britain where distant relatives or people from the same village or district live as lodgers. They are temporary members and leave to establish their own households when their wives and children join them.....this situation is changing as more Pakistanis are bringing over their families to this country. Similarly, young men travel to Pakistan to get married, and return to establish separate households (Anwar, 1985, pp.54-55)."

Anwar cites the following examples (fig.2.2) of joint households in Britain from his study of British Pakistanis in Rochdale.

Nine of the eleven intensively studied families in this research lived as nuclear families. The other two families went through stages of living as a nuclear and extended family as family circumstances and finances dictated.

### The Kinship System

Beyond the household it is the kinship system which regulates and structures relationships. Alavi (1972) states that:

"Underlying much of the literature on South Asian rural societies is the assumption that these societies are structurally similar, if not identical in every detail, and that the distinguishing feature of the structure of social institutions in those societies is their focus on caste and the related jajmani system as bases of social organisation. The kinship system, which is generally accorded a central place in social anthropological analyses is relegated to the background. In the muslim rural society of the West Punjab, however, the contrary is true. There it is the kinship system rather than caste which embodies the primordial loyalties which structure its social organisation. The basic institution of the kinship system of Muslims of West Punjab is the "biradheri" (p.1)."

The biradheri (meaning "brotherhood") is an endogamous group whose members claim descent in the paternal line from a common male ancestor, but in certain contexts the word is used to refer to individuals or groups with whom there is a brotherly and hence loyal relationship. Anwar (1985) defines the nature of biradheri as the following:

"The descent group Biradheri includes all those who claim and can trace links of a common paternal line. As Pakistani society is patrilineal all the inheritance is through the male line and so is the Biradheri. Daughters belong to the Biradheri of their father but after marriage are also included in the Biradheri of their husbands. It is often difficult to draw the line

where the Biradheri ends, as this depends on the person defining those he thinks belong to his Biradheri. In practice it depends on the contact maintained with each other, the degree to which endogamy is practised, the frequency with which members meet to take decisions which affect the Biradheri as a whole, or participate in ritual ceremonies and in Vartan Bhanji, an exchange of gifts and a whole range of services. Distantly related consanguines, those related affinally, or friends who have assumed either affiliation due to regular rituals or certain kinds of exchange of services may be incorporated into one's Biradheri. This type is usually called "fraternal Biradheri (p.63)".

Respect is ascribed to biradheri elders who have the power to reprimand deviants and ensure the cohesion of the group.

Related to the effective functioning of the biradheri is the practice of "vartan bhanji" which is a mechanism of gift exchange, denoting affiliation with those participating in the exchange. The term vartan bhanji literally means "dealing in sweets" but in social terms means "dealing in relationships". It involves the exchange of sweetmeats, gifts and includes the exchange of services and favours. The transactions usually take place on occasions such as births, the circumcision of a son, visiting relatives and marriage. Gifts may also be given on other occasions such as departure for a pilgrimage to Mecca and in the British context, on departure to or from Pakistan. An example of vartan bhanji as a favour would be the offering of an interest-free loan to someone for the deposit on a house, or to help with the cost of a marriage. Alavi (1972) states that the exchanges of gifts are:

"....transactions in a perpetual cycle of regular

reversal of ritual debts between the households. Each payment, notionally, consists of two parts; one part extinguishes the pre-existing debt and another part creates a new debt in the reverse direction (p.11)."

The biradheri functions as a welfare, financial and advice service. The system of gift exchange gives financial support to biradheri families in times of considerable expenditure and expresses official alignment and solidarity with other households. Interaction between biradheri members, however, is not always smooth and there can be serious and prolonged conflict.

A wider and much looser network of affiliation than biradheri is the "quom", which often has relevance in terms of choosing marriage partners:

"Although the caste system is rejected in Islam there are clear vestiges of the pre-partition social structure. Notions of purity and pollution, restricted commonsality and certain other features of the caste system are less evident in Mirpur and Pakistan but there is a general hierarchy of castes (with landowning castes at the top and service castes lower on the scale). Not all sons follow their father's traditional occupation (e.g. "Lohar"-blacksmith, "Nai"-barber, etc.) but the caste ("quom") name is retained and is thus significant in certain situations (Saifullah-Khan, 1977, p.60)."

The biradheri extends beyond the village to other villages in the locality and closely related biradheri members who live at long distances remain in close touch and attend important family functions. Fellow villagers, although often unrelated are another category with whom a household may have close

ties. Biradheri networks also extend outside of Pakistan taking in members of their biradheri in Britain and in other countries where Pakistanis have settled. Several commentators (Anwar, 1985, Dahya, 1973, Ballard, 1988 Saifullah-Khan, 1974) state that to have a complete picture of the functioning of the Pakistani population overseas it is necessary to regard the whole as a single system of socioeconomic relationships (see fig. 2.3).

The institutions of biradheri and vartan bhanji are firmly entrenched in the social fabric of Pakistani society whether in rural or urban Pakistan or in Britain. In each setting it is in virtually identical ways that affiliations are made and solidarity among kin maintained to form networks within which obligations, support and control operate.

#### The influence of kinship.

The first generation British Pakistanis followed a specific pattern of behaviour which ensured the continuation of the familiar, supportive institutions and values of life in their native country. It also illustrates the tight network within which they operated in Pakistan and which continued to be a part of their lives despite the distance.

The decision to migrate was made by family and kin and those immediately involved financed the journey. The head of the household, or of the immediate biradheri grouping, selected

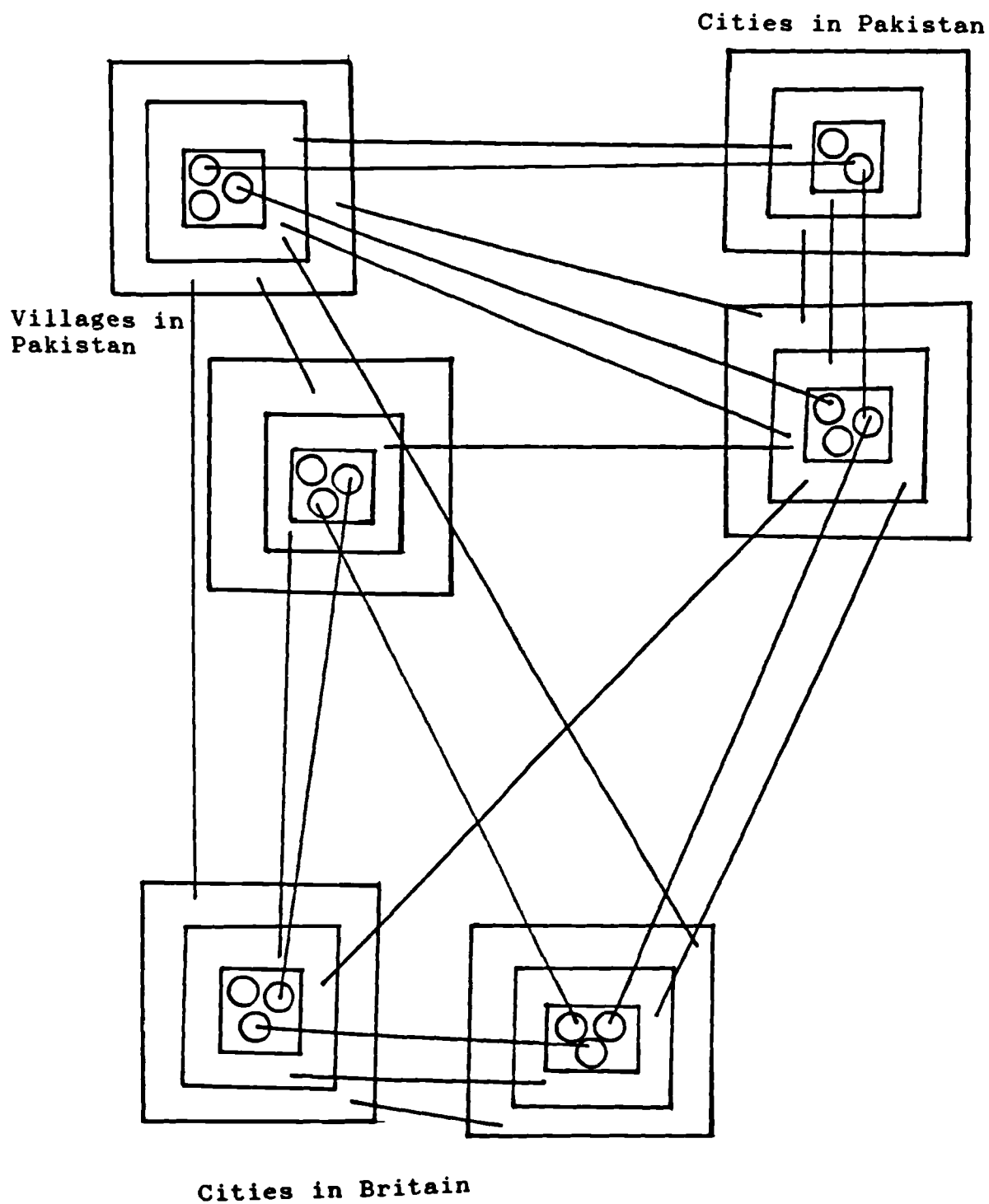
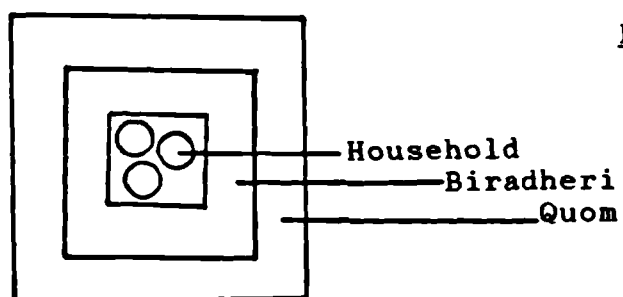


Fig. 2.3: Social networks



the migrant and made the preparations. The joint decision reinforced the ties with kin and community, thus cementing feelings of affection and determination.

Whilst in Britain the contact with the family in Pakistan continued. Dahya (1973) provides a description of the attitudes and lifestyles of the first-generation migrants living in all-male "dormitories" whose main reason for migration was to improve the family's corporate resources, whose emotional ties remained in the village and for whom all aspects of life in Britain were transient. He cites the following comments to him as typical of those made by male migrants at the time

"Will the English people think better of me if I live in a modern house here? Better to build a pakka (brick-built) house in the village where people matter." "Let them know (the people in England) how we live in style and comfort." "Show them photographs so that they should not judge us by what they see of our houses in Sparkbrook (p.255)."

Both Anwar (1985) and Dahya (1973) refer to the "myth of return" which characterised the thoughts of migrants. This notion of eventual return, fed by the head of the household in the village and by the migrant formed the base of a life pattern on which decisions were made and joint family responsibilities carried out which secured the family's future and provided a focus for the migrant in terms of his base, status and future. However,

"The cycle of moves and constraints do not end with the death of the family head. The migrant himself returns



home on the death of the head, leaving his younger brothers and sons behind in Britain. As the head of the family he begins to exercise constraints on the rest of his kinsmen in Britain just as his father did during his sojourn in Britain (p.257)."

The initial decision to leave wives and children in the village fitted neatly into the overall plan, the kinship network and consanguineous marriage pattern ensuring that they were in protective surroundings. Several factors, mentioned earlier, converged to change the nature of Pakistani settlement from being temporary to more permanent, so that by the mid 1960s and early 1970s there were settlements of Pakistanis in British cities comprising kin which was an extension of kin at home. Mosques and other facilities such as halal butchers and Pakistani grocery shops were established, surroundings thought to be far more conducive for women and children wherein there was a possibility of maintaining the same moral values as in Pakistan. By this time men had also absolved themselves of some of the financial responsibility to their extended family. Not least was the desire to be united on a more permanent basis with their wives and children, whilst continuing to work in Britain within a network of kin and surroundings, now more familiar than those in Pakistan. Although links are still maintained with Pakistan and many may still harbour a desire to return to their native country, a definite shift in attitude towards residence in Britain has taken place, which more than likely, is intensified among the British-born Pakistanis.

Not all British Pakistanis, however, have followed the path of the majority. There are individuals who, for various reasons, have not maintained ties with their homeland, and some who have broken their ties even with kin in Britain.

SOCIAL INTERACTION

"Relationships in the village are based on personal, face-to-face interaction. Village life is outdoor and gregarious. Everyone knows everyone else and the village forms a moral arena in which reputations are assessed and reassessed, and potential deviants pulled back into line. Child rearing is relaxed and shared, and children learn by observation rather than by training. They receive a lot of affection and assume responsibilities toward different kin and villagers at a very early age. The distinct patterns of behaviour between the sexes is established by the time of puberty (Saifullah-Khan, 1977, p.62)."

Family or biradheri elders are highly respected and often referred to as "our bozorg" (our elders). The term is applied to both males and females, denoting a position of respect for the person. Much of the literature on kinship among Pakistanis stops short at describing the system as patrilineal with final authority resting with the male head of the household. However, it is important to note the processes prior to the final decision in which consultation with other members of the household takes place, particularly with the female elders. Final authority and responsibility rests with the eldest male only in the male domain. In interaction with female members of their own and other households it is the women who have ultimate responsibility in initiating and maintaining cordial relations. The female role is virtually indispensable in the arranging of marriages (see pp. 133-135).

If there are conflicts over any action to be taken then it is not uncommon for men and women to voice opinions and negotiate

as the kinship system and the multiplicity of relationships created by consanguineous marriage provide support structures for both men and women. There are however rules of decorum which are expected to be observed. A woman is not meant to undermine a male's authority in his domain and similarly a male will be reprimanded by a woman for interfering in her domain. The importance of such subtleties in male-female interaction are possibly underestimated in the literature, as much previous work has been carried out either by male researchers who do not have access to the female world or, by non-Pakistani females who have access to the female world, but only partial access to the male world. Hence, during participant observation, no matter how much rapport is established with a respected and loved outsider, nevertheless, on a subtle level they remain an outsider with whom a public face is maintained. I draw this latter point from my own personal experience of being "observed" as a British Pakistani, and through comments from British Pakistanis involved in this research who have said, " I'll discuss this aspect of family life with you, there's no point in hiding it, you're one of us. You know these things happen. It may have happened in your family too." It is only by researchers "of the system", and aware of the need for objectivity that a greater depth in research in the interpersonal sphere can be achieved. Even male members of a household, including male Pakistani researchers, would not necessarily be aware, or be made aware of the processes and interpersonal dynamics

involved and practiced by women to maintain the smooth running of a household or kin network. Female members have far greater involvement with and discussion around the interpersonal skills needed to maintain good relations in the extended family in which they play a central role. As female interaction is informal, subjective and entails frequent contact, subtle strategies for maintaining control over their personal sphere whilst still maintaining cordial relations are taught verbally and non-verbally during socialisation. It is therefore incorrect to assume that it is only males who exercise power and makes decisions.

The conventional division of labour between men and women is practiced. Men are the main providers and if farmers, work in the fields, whilst women look after the home, the children and the livestock. During busy periods women also participate in agricultural and farming activities. Due to the outdoor nature of village life there is considerable interaction between men and women. The degree and nature of the interaction is, however, affected by muslim beliefs about behaviour between the sexes. In all muslim societies a certain degree of distance and formality in public between males and females is believed to be healthy. It is perceived to be an acknowledgement of sexual dynamics between the sexes with formality in behaviour serving as a control mechanism. The formality between the sexes takes various forms, for example, there are usually two sitting rooms in a home so that

men and women can have separate areas, particularly if unfamiliar people visit, whether male or female; avoidance of familiarity with members of the opposite sex who are not well known; observance of rules of dress (covering) in the company of the opposite sex. There is little segregation with members of one's immediate, extended or joint family.

Saifullah-Khan (1976a, pp.103-105) notes that there is increased segregation of men and women in cities in Pakistan and in Britain, as the urban setting often means that one's neighbours and service providers are people with whom familiarity has not developed over a long period. She goes on to say that with increased wealth in an urban setting more space is available in the home to allow segregation while women are no longer required to work in the fields; both factors serve to reflect the prestige of the family.

Whilst acknowledging the existence of sexual dynamics between men and women each individual is expected to accord the other respect and the men and women of one's village are sometimes referred to as one's brothers and sisters meaning that a sense of mutual respect and responsibility is expected. Men as well as women are expected to be suitably attired in each other's presence. With men and women who are strangers, that is, people with whom there is minimal contact, to be formal in one's behaviour is to be self-respecting and to accord respect to the other. This applies as much to men as to women. If a

couple are together and meet a man not well-known to wife, then the husband is expected to do the talking. Conversely, if they meet a woman the woman would do the talking. If they meet someone they both know all of them would converse. It is these dynamics that are taking place in the G.P. surgery in Britain: When a husband mainly converses with a male G.P. and the woman remains quiet, it does not mean she is being submissive, but simply following rules of etiquette. It does not follow that the husband necessarily makes the decisions. In Britain, however, the situation is complicated because most women are unable to participate due to the language barrier, whereas adult women are not chaperoned by men when visiting a doctor in Pakistan. They may, however, ask or expect one to accompany them.

In the cities in Pakistan dress and behaviour patterns are modified due to an urban setting but male-female interaction in the cities is based on the same principles. Further modifications take place in Britain, but the basic principles of interaction remain the same.

MARRIAGE

Culturally and religiously all men and women are expected to marry. Most marriages in rural and urban Pakistan and among British Pakistanis take place within the extended family or biradheri (see pp.278-288 for a discussion of consanguineous marriage). The norm is for parents and adult siblings, particularly the mother and sisters, to choose a partner for the individuals concerned based on their personalities and ages, as well as the relationship between their parents. The marriage is viewed not only as a relationship between two individuals, but the creation or further consolidation of a bond within the family or between two families. For most Pakistani men and women it is as alien to be expected to seek one's own marital partner as it is for people of Western cultures to ask their family to arrange their marriage.

Marriage between two families, related or unrelated, is only contemplated if respect exists between them and a desire to further consolidate existing bonds. By the same token, it is expected that the couple concerned will also be respected and loved among their respective in-laws. The various personal and family implications of a match are discussed beforehand and only if these are satisfactory, will a marriage proposal be made. The proposal is made by the mother of the man concerned, who visits the mother of the woman, who then discusses it with her husband and family before giving the final decision. The mother of the man is usually accompanied



by another adult female member of the family. The emphasis on family and one reason for the preference for close-kin marriage derives from the continuing rights and obligations that Pakistanis have towards their kin. So, once a proposal has been made between close kin there is an expectation that it will be accepted. A rejection of such a proposal without an understandable reason is taken to be a slight against the family. If there are females of marriageable age in an extended family it is expected that the parents of suitable males will ask for their hand in marriage. If this does not happen and a marriage is arranged outside of the family, it is regarded as dishonouring one's obligations to one's family and will affect the future interaction of the families concerned, sometimes quite openly and sometimes only on a subtle level.

In the case of a distantly-related family, biradheri members with whom there has not been much contact or unrelated families a woman known to both families acts as a go-between, furnishing each family with details of the other and of the prospective bride and groom, before an initial visit or proposal is made. If the initial enquiries show the match to be unsuitable then any further steps are curtailed.

Most marriages in Pakistani cities and in Britain follow the same conventions as in the villages, with some modifications. In an urban setting there are usually more opportunities for males and females to meet individually (particularly in

educational establishments) and hence there may be a tendency for more marriages to be based on individual rather than family considerations.

Since close-kin marriage is very much favoured, residents of different villages, cities and countries are connected to members of the same family or biradheri in other villages, cities and countries by a multitude of ties of kinship, marriage and affinity.

Although consanguinious marriage is favoured in muslim societies, it is neither encouraged or discouraged in the Qur'anic text. When consanguineous marriage patterns have been discussed with the families in this study their response has been that neither the Qur'an nor the Prophet Muhammad discouraged the marital unions practised. If those unions had been detrimental for any reason then they would have been included in the marital unions that are prohibited. The following verses from the Qur'an state the relationships between which marriage, and through which, procreation can take place:

"Forbidden to you (in marriage) are your mothers, your own daughters, your sisters, your aunts on your father's side and your brother's and your sister's daughters, your foster mothers and your foster sisters, your mothers-in-law and step daughters who are under your guardianship (since their mothers are wives of yours with whom you have consumated marriage; however if you have not consumated it with them, it will not be held against you) and wives of your sons who are your own flesh and blood; nor may you bring two sisters together (under one roof), unless this is a thing of the past (Qur'an, Surah 4, verse 23)."

ISLAM

Western images of Islam are predominantly of a dogmatic, doctrinaire religion with a hierarchical structure of imams, who are viewed as powerful figures in the community. Couples in this study, however, made their own decisions based on their understanding of their faith, in which they found much solace. Their religious beliefs played a large part in helping them to cope with the demands of a chronically sick child in the family. In the following section I have attempted to explain the nature of Islam in a manner that goes beyond just a statement of the basic tenets of the faith, in order to illustrate some aspects of the faith and its followers that are not immediately apparent, but which play a significant role in the spiritual, emotional and psychological make-up of many, though not all, British Pakistani muslims. In particular it is necessary to clarify the role of the "pir"<sup>1</sup> in Pakistani life, since pirs play a significant part in the ways that the families in the study have sought to deal with their child's chronic disease.

Islam is the faith of the Muslims, who number 900 million worldwide. It is a monotheistic faith which stands in the Abrahamic tradition, preceeded by Judaism and Christianity. Presently there are two major sects in Islam, Sunni and Shi'a, and a number of schools of thought. Although there are some differences between the sects in terms of

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<sup>1</sup>spiritual guide.

interpretation and practice of some aspects of the faith, its basic principles are adhered to by both sects. The Shi'as are a minority in the Islamic world, but form the majority population in Iran and some Middle Eastern countries. In Pakistan the vast majority of people belong to the Sunni sect, though there is also a sizeable Shi'a population. The Qur'an, the sacred book of Islam is believed to be the direct word of God revealed to the prophet Muhammad who lived between 570 and 630 A.D..

Frithof Schuon (1989, p.13), has described Islam as a meeting between God and man. From an Islamic perspective, Islam views God as the essence of the cosmic Reality (encompassing that which is visible and apparent and that which is invisible and not apparent), the Omnipotent and the Absolute. Man is viewed as:

"A theomorphic being endowed with an intelligence capable of conceiving the Absolute and with a will capable of choosing what leads to the Absolute (Schuon, 1989, p.13)."

The Qur'an refers to Islam as a "dheen" (a way of life). It addresses the whole of mankind, women as well as men, and emphasises an individual's link with God, and responsibility to other human beings as well as to the society in which he or she lives. The family unit and nurturance within that unit has a particularly important place in the social fabric of societies practising Islam. Jansen (1979) has succinctly summed up his understanding of the faith:

"It is a total and unified way of life, both religious and secular; it is a set of beliefs and a way of worship; it is a vast and integrated system of law; it is a culture and civilisation; it is an economic system and a way of doing business; it is a polity and a method of governance; it is a special sort of society and a way of running a family; it prescribes for inheritance and divorce, dress and etiquette, food and personal hygiene. It is a spiritual and human totality, this-worldly and other-worldly (p.17)."

Muslims view life on Earth as one stage in the total existence of human beings, with another realm of existence after death and the Day of Judgement. The purpose of this life is to realise one's full potential in a righteous manner. In order to realise individual potential both temporally and spiritually, a code of conduct is prescribed for Muslims which is based on Qur'anic teachings, "hadith" (sayings of the Prophet) and the "sunna" (actions of the Prophet).

Worship, in Islam, is a broad concept and encompasses a whole range of private devotions, social actions and human relations (McDermott and Ahsan, 1980, p.23). For anyone choosing Islam as their way of life, there are five essentials of worship enjoined in the shari'at (the Islamic code of conduct), which are stepping stones towards spiritual and temporal fulfillment. Shaykh Abd al-Qadir as-Sufi ad-Darqawi (1979) states the importance of these injunctions and of the shari'at as a whole:

"The shari'at of Islam is the confirmation that there is no divinity but Allah and that Muhammad is the messenger of Allah. It is to pray five times a day the ritual prostrations. It is to fast the month of

Ramadan. It is to pay the zakat tax of wealth. It is to take, if possible, the Hajj to the pure House of Allah and the plain of Arafat<sup>2</sup>. It is based on these and confirms that the one following the shari'at has elected to live within the broad moral parameters set down in the Qur'anic commands and according to the guidance of the sunna, the life pattern of Muhammad, blessings of Allah and peace be upon him. Having accepted the shari'at is the deep cognition that the human creature is limited, is in a body, and thus like all bodies in the physical world obeys given laws. There is no compulsion in the life-transaction, thus it cannot be called "organised" religion - no - it is the self chosen pattern of life one has adopted in order to deepen knowledge untill one reaches one's own source, one's spring of life, to drink of the water of illumination (p.2)."

The above quote is taken from the writings of a sufi master. It represents the thought pattern of one who belongs to one of the two categories of learned followers in the Islamic tradition, the sufis. The other category is that of the "ulema" (scholars).

"In the course of time, Islam's scholars and doctors of religion came to comprise two main categories. One of these continued to be the "ulema" (scholars), and their primary task remained religious study, teaching and writing. The other category came to be known as sufis (mystics) or "swaliheen" (the pious ones). These latter were also basically religious scholars; indeed in classical sufism<sup>3</sup>, no one who was not well-versed in the Prophet's shariah<sup>4</sup> (temporal code) could aspire or claim to be a sufi. With the great stress thst Islam places on the acquisition of knowledge, religious as well as secular, sufism could scarcely hope either to inspire confidence or to acheive its lofty ends without a solid scholarly base. However, the sufis projected and propogated Islam primarily by practical example

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<sup>2</sup> The annual pilgrimage to Mecca in Saudi Arabia.

<sup>3</sup> In Arabic, sufism (or Islamic mysticism) is termed "tasawwuf", hence the word "sufi" from which the anglicised word sufism is derived.

<sup>4</sup> The Arabic word, shari'at, is spelt in a number of ways by different authors.

rather than by teaching or theoretical exposition alone. In teaching too, they emphasised the spiritual aspects, so as to promote the purification of the self.....(Khan, 1980, p.4)."

The sufis have had a tremendous influence in shaping the nature and practice of Islam in the Indo-Pak subcontinent, where Islam's emphasis on egalitarianism was in direct contrast to the rigid hierarchy of the Hindu caste system and was particularly attractive to the low-status castes. Khan (1980), amongst others, questions the validity of the opinion that Islam was spread primarily through military conquest. He states that:

"The actual task of bringing it to the hearts and souls of the new millions was performed by the ulema and sufis, many of whom served in and accompanied the Muslim armies in their campaigns, and many also came as traders and craftsmen. Islam exists today in some countries of the Far East which were never militarily "conquered" by the Muslims (p.9)."

Khan cites the Encyclopaedia Britannica (1974) as stating that:

"The main contribution of the sufi orders.....is their missionary activity.....Sufism has helped to shape large parts of Muslim society.....The missionary activities of the sufis have enlarged the fold of the faithful. The importance of sufism for spiritual education, and inculcation in the faithful of virtues of trust in God, piety, faith in God's love, and veneration of the Prophet cannot be over-rated (Vol.9,pp 947-48)."

The Pakistan Tourism Board lists 86 large and 37 smaller shrines of Muslim sufis in Pakistan. These shrines, especially the major ones, are a hub of activity, particularly on Fridays, the Muslim sabbath, and on the

annual "urs" (day of death of the mystic). The urs often spreads over three to five days and may be declared a local or regional holiday, often assuming the character of a "mela" (fair). A visit to a shrine reveals it be a microcosm of life in Pakistan. The rich, the poor, the healthy, the sick, men, women and children are found there. It is a place where food and shelter is constantly available and which belongs to no-one but at the same time belongs to everyone. Some come simply to offer prayers for the departed mystic whose writings have influenced them, others come for spiritual sustenance and to soak in the devotional atmosphere, others come with specific difficulties in their lives which they want resolved and to ask for blessings for the resolution of problems. It is believed that the good works of the mystics have made their shrines a hallowed place and that the blessings of God are in abundance in a place where constant prayer takes place.

There are a number of sufic orders into which someone wishing to develop his or her spirituality can be initiated. The "murshid" (spiritual guide) is the one who has enough knowledge and has developed his or her spiritual awareness to a degree which enables him to guide a "murid" (disciple) along the same spiritual path. The initiation into an order is called taking "bai'at" (oath of allegiance) from a chosen murshid. Most, but not all, of the well-known murshids have



been men, whereas both men and women are known to be murids<sup>5</sup>. I refer here to the "known" sufis, for as Shah (1968) states:

"The sufis who have been publicly known are only a minority of the total of the sufis: those who could not keep out of prominence (p.312)."

The murshid's consent to guide a person must be sought to consolidate the relationship. There are no examinations in this search for knowledge. It is a purely mystical journey, with its roots embedded in a continuance of everyday life and obligations, though the murshid may suggest periods of "dhikr"<sup>6</sup> in solitude. Before a disciple can take on the role of being a spiritual guide to others, permission must be sought from his or her own murshid. The descriptives murshid and murid are used for those who have formalised their spiritual relationship. A person who is a murshid may also be referred to as one's "pir" (also meaning spiritual guide) by someone who seeks advice but who has not entered into any formal relationship for the purpose of spiritual advancement.

The shrine is also a place where people come to pirs to ask for advice for specific problems. As the family studies in chapter 12 show, many women visited their pirs to seek advice on whether a termination of pregnancy was permitted given

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<sup>5</sup> Eaton, (1978), p.87. Referred to in Lewis, P. (1985), p.70.

<sup>6</sup> Remembrance of God.

their risk of a thalassaemic child, or to ask for blessings, amulets and for prayers to recite to be granted a healthy child. Some of the women made journeys to Pakistan to visit a family pir. Others visited pirs either resident in England or who were visiting England. Lewis (1985) cites the importance of the particular nature of life at a shrine for women who visit them:

"As a murid, or simply as a visitor to a shrine, a woman often entered a wider and more spacious world: here she could enjoy the aesthetic satisfaction and emotional release of devotional songs in the vernacular: here she could bring her problems, illnesses and anxieties and be guaranteed a sympathetic ear; here she could participate in a rich cycle of festivals, religious and secular (p.71)."

As indicated earlier, pirs also travel to different countries, including England. Some may be resident here with their group of followers and disciples. News of their visits is usually passed along the grapevine for those who would be interested in the discourses. Such meetings are usually held in people's homes. One of the families in this study had a room specifically set aside for these sorts of gatherings. Less learned, but knowledgable men also travel around and visit villages and cities in Pakistan or cities in other countries imparting information, knowledge and wisdom. Such a man is not attached to any particular mosque though he may be of a particular sect or order, in which case the people choosing to listen to him or meet him may be predominantly those with the same sect affiliations. Among people of urban background there is no compulsion to attend meetings or

shrines or to believe in the wisdom of pirs. However, Chaudrey (1979) states that in village society it is believed that someone without a pir "does not have a source of spiritual guidance and his sins cannot be written off on the day of judgement (p.68)."

The sufic way is very much a living tradition both in Pakistan and in Britain. The followers of the tradition are not obvious as there is no recognisable formal structure or dress. The writings of the earlier sufis have, however, permeated the literature and music of Pakistan; these arts are also available in Britain. Evidence of the teachings, with its stress on living a righteous life and percieving total existence as encompassing an after-life and the unseen, have formed some of the popular sayings in Pakistani culture.

Visiting shrines and the people associated with them, however, is a contentious issue among Muslims, though this is not an attack on the sufic tradition. Some denominations of Islam and individuals hold the view that people visit shrines to ask the deceased mystic to answer their prayers and thereby perform "shirk"<sup>7</sup>. It is also known among Pakistanis and British Pakistanis that there are men who project themselves as pirs, and who are believed to be such, but who have limited knowledge of Islam and charge for their advice,

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<sup>7</sup> Associating another with God, thereby compromising His Oneness, the antithesis of a fundamental Islamic belief.

either directly or indirectly.

The practice of Islam varies from country to country, from sect to sect, from city to village, but there is little debate on the fundamental beliefs or the major injunctions of the shari'at. Local practice of the faith is often tinged with the cultural practices of the region, leading to some confusion between cultural and religious practices. In Pakistan the days are punctuated by the five prayers and the year by the religious festivals and the month of Ramadan. Though not everyone prays five times or is mindful of religious traditions but it is widely acknowledged that these are respected practices.

#### Mosques and imams of mosques

Mosques are one of the focal points in every Muslim community, particularly for the men, in Pakistan and in Britain. The imam of the mosque leads the congregational prayers on Friday when he usually also delivers a "khutba" (sermon). He also looks after the affairs of the mosque. In Britain women do not usually pray in a mosque, although in Pakistan some do go to pray or listen to the khutba. The activity surrounding a mosque is largely dependant on the personality, religious knowledge and experience of the imam as well as the standing he has in the local community. Hence, a mosque can be a place solely to meet and perform

communal prayers or it can be a centre of learning, debate, support and even political activity. In Britain, where recitation of the Qur'an and Islam is not taught in schools, it is often taught in the mosque after school hours. Sometimes, the wife or daughter of the imam may teach recitation of the Qur'an to very young girls and boys.

The imam is not required to go through formal religious training. The main pre-requisite for being an Imam is being of worthy character and commanding respect by virtue of piety, among the local community. Whether his counsel is sought is largely dependant on his personality, but to have a pastoral role in the community is not an automatic expectation of his role.

#### Islam and termination of pregnancy

The popular belief about the termination of pregnancy among Muslims is that it is not permissible unless the mother's life is in danger. Discussions with learned Muslims revealed that there were different schools of thought on the subject, some pronounced it permissible before three months gestation and others before ten weeks gestation on the grounds that the "ruh" (spirit) enters the fetus at this time, whilst others said it was not permissible at any stage unless the mother's life was in danger. The limits of time did not allow a detailed study of this issue, which would require

considerable research. There was a general consensus that a fatwa<sup>8</sup> had not yet been passed on termination of pregnancy in the context of chronic disease. As the family studies show the couples in the study held the popular Islamic view on terminating a pregnancy, but in the light of their situation, made their own decisions.

Termination of pregnancy is not legal in Pakistan, unless the pregnancy endangers the life of the mother. However, illegal abortions are performed.

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<sup>8</sup> "A legal opinion given by a "mufti"- in contrast to a decision ("hukm") given by a judge ("qazi")- on the basis of Islamic law in answer to a question, commonly concerning family law and inheritance (Lewis, 1985, p.106)."

## THE BRITISH PAKISTANI POPULATION

### Population structure

It is estimated that presently there are approximately 385,000 people of Pakistani origin living in Britain of whom nearly half have been born in Britain; in 1984 almost 45% of British Pakistanis were born in Britain (table 2.1).

Virtually all British Pakistanis are British nationals. The decision to take up British nationality was eased by the Pakistani Government allowing Pakistanis with British nationality to retain their Pakistani citizenship.

All persons born in UK	118,252
All persons born outside UK	177,209
Total persons, 1981 estimates	295,461
Total persons, 1987 estimates	384,320

Table 2.1<sup>1</sup>: Persons resident in private households with the head of household born in Pakistan

The age distribution of British Pakistanis is shown in fig. 2.1. People now in their forties and fifties came to Britain as young adults in the 1950s and 1960s. People currently of reproductive age in their mid-twenties and early thirties were mostly born in Pakistan and came over later as dependants. The bulk of the people under twenty are UK born. As Anwar (1985) notes the British Pakistani population is younger than

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<sup>1</sup> Source: Anwar, M. (1985), p.249.

the indigenous population. He estimates that 40% of British Pakistanis are under 16 years of age, compared with 22 percent of the general population. There are relatively few elderly people but due to the young population structure many women are of reproductive age and hence there is a higher number of births plus a larger final family size than in the general population.

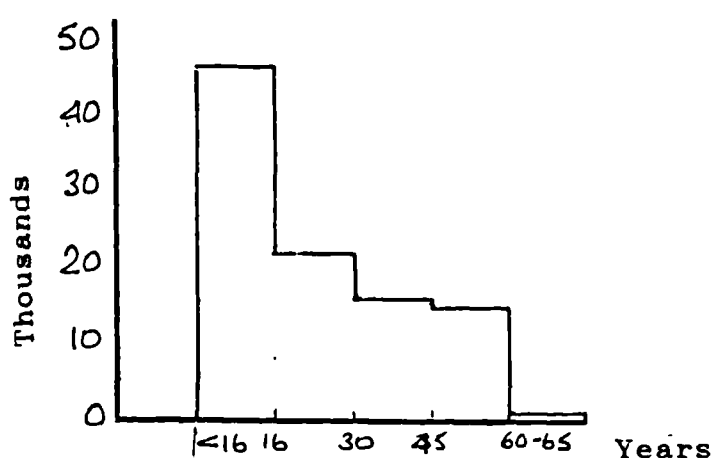


Fig. 2.4<sup>2</sup>: Age distribution of British Pakistanis, 1983

#### Geographical distribution

British Pakistanis reside in certain areas of Britain rather than being dispersed throughout the country, as illustrated in map 2.1. (A detailed breakdown of the areas of settlement can be found in appendix 1). There is a large concentration of British Pakistanis in the Greater London area, the West Midlands, West Yorkshire, Lancashire and Central Clydeside, mainly in Glasgow. Thirty one per cent of British Pakistanis

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<sup>2</sup> Source: Labour Force Survey, (1984), table 5.5.



live in London and the South East, twenty two per cent in the conurbations of the West Midlands, twenty one per cent in Yorkshire and Humberside , and sixteen per cent in the North-West. The last areas listed really make a single loose aggregate of 36% (Anwar, 1985, p.251<sup>3</sup>). Hence, these are the areas where genetic health services appropriate for the population of Pakistani origin need to be concentrated.

city which consist predominantly of one particular ethnic group from that migrant population. Areas where mainly people of Pakistani, Indian or Cypriot origin and so on have settled have become associated with that population, i.e., Bradford is known as a Pakistani city, Leicester, a Gujarati city, Southall, a Sikh area and Haringey, a Cypriot borough.

"Extensive research shows that individuals with similar cultural origins tend to cluster together and thus become residentially segregated from the rest of society (Lieberson, 1961; Jones, 1967; Richmond, 1972). During the first stages of settling in a new society residential proximity enhances interaction and helps to ease culture shock (Jones, op.cit., p.4) It allows people with similar values to maintain their group norms, to preserve a sense of ethnic identity and to feel secure in a familiar social network (Richmond, op.cit. p.4). People with similar and specific needs tend to cluster together in order to establish institutions catering for those needs. This pattern applies to Pakistanis in Britain (Anwar, 1985, p.11)."

In addition to the above, a group of people need to reach a critical size so that they can begin to have their own facilities, such as mosques and halal butchers, at hand. However, whilst kinship networks played a major role in the establishment of clusters of British Pakistani settlements in the inner city areas, wider discriminatory political, economic and social forces within the host community have also influenced the settlement pattern. The same forces are still a significant factor in perpetuating the employment and housing of British Pakistanis, along with that of other ethnic minorities (see Deakin 1970, Brown, 1984).

A British Pakistani area in an industrial city is usually made

up of rows of terraced houses, leading off from one major road in the city where the shops, restaurants, banks, travel agents and so on cater predominantly for British Pakistani tastes and needs. If there is no purpose-built mosque nearby, one or more large houses will have been purchased by the community and used as a mosque for prayers, religious gatherings and for teaching recitation of the Qur'an to children. In one city alone there may be a number of mosques. In the neighbourhoods families will more than likely have relatives or kin living in the nearby streets. Men, women and children will make visits during the day for mutual support, advice or just company and children play on the street. Women can be seen visiting the local shops, on foot, sometimes with their small children. Often, an elderly or particularly religious woman takes the responsibility for teaching the children recitation of the Qur'an after school hours. If a woman can sew and needs the extra money she may become the seamstress for the other neighbourhood women. Some shops may be focal points for information and interaction, particularly for the men. So the area consists of clusters of families interconnected by national, village or kin ties where support, particularly in times of distress, is at one's doorstep but where competition and conflict also exists. To some extent it is a reconstruction of the societal set-up in the village.

### Women's employment

For the majority of British Pakistani women life centres around the home and family. Seventy-eight per cent of Muslim women in Britain of Asian origin (mostly of Pakistani origin) are housewives. They provide the lowest proportion of all women in the labour market (table 2.2).

The highest proportion (43%) of economically active British Pakistani women work in non-manual jobs (table 2.3). Twenty-two per cent are in semi-skilled manual jobs and only two per cent are doing unskilled manual work. Of working Muslim women the largest percentage (42%) are aged sixteen to nineteen years (table 2.4), that is, the group includes few mothers with children. Probably they are second generation British Pakistanis with a good command of the English language and an awareness of the functionings of British society. Given that "economic activity among Asian women is related very strongly to fluency in English" and that 70% of adult British Pakistani women speak English "slightly" or "not at all" (Brown, 1984, pp. 151 and 137), the low uptake rate of employment probably reflects the fact that most of the jobs available to the majority of the women are low paid and probably with bad working conditions.



There is an increasing association between homeworking and ethnic minority women but reliable figures on the extent of homeworking<sup>8</sup> amongst women are not available (Allen and Wolkowitz, 1987, p.13). Though a specific enquiry about the extent of homeworking was not made in this study, one of the mothers in the eleven intensively studied families, whose husband was unemployed was doing homework. Anwar (1985) states that homeworking among British Pakistani women is a compromise between socio-cultural factors and the need to earn money (p.133) whereas Allen and Wolkowitz, referring to British Pakistani homeworkers, argue that "The determination of their work options was clearly more complex than popular myths regarding the housebound "Asian" women imply." They found that most British Pakistani homeworkers

"cited material and ideological constraints similar to those faced by the other homeworkers. Like white homeworkers, the Pakistani homeworkers usually mentioned the need to care for children. All worked out of

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<sup>7</sup> Source: Brown, C. (1984), p.187.

<sup>8</sup> "Homeworking is the supply of work to be performed in domestic premises, usually for piecework payment."

economic need and, like other homeworkers, contributed a substantial amount to household earnings (p.82)."

### Men's employment

The most recent comprehensive figures on economic activity in Britain are to be found in the 1981 census. In 1981 82,860 British Pakistanis (76,930 males and 5,930 females) were economically active; 93 per cent of all economically active British Pakistanis were male (Anwar 1985, p.97). British Pakistanis were more likely to be employed as labourers in textiles, engineering and allied trades than males from other groups and less likely to be working in administration, management and sales. They are concentrated in the manufacturing industries (Third PSI Survey 1984, pp.157-159). In 1984 over 19% worked in textiles, compared to 2.5% of all economically active persons and 16% worked in metal manufacture and the metal goods industry. Along with other ethnic minority workers the occupations had low pay. The job levels of all employees by ethnic origin in 1984 are shown in table 2.5<sup>9</sup>.

Unemployment rates are higher among all ethnic minorities than among the white population (Brown, 1984, pp.151-155). In the present economic recession unemployment among British Pakistanis has increased along with that in the British population as a whole. As most British Pakistanis were

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<sup>9</sup> Source: Brown (1984), p.197.

Job Level	Column percentages								
	White	West Indian	Asian	Indian	Paki-stani	Bangla-desi	African Asian	Muslim	Hindu Sikh
Professional, Employer, Management	19	5	13	11	10	10	22	11	20
Other non-manual	23	10	13	13	8	7	21	8	26
Skilled Manual and Foreman	42	48	33	34	39	13	31	33	20
Semi-skilled Manual	13	26	34	36	35	57	22	39	28
Unskilled Manual	3	9	6	5	8	12	3	8	3
									6

**Table 2.5: Job levels of men: all employees by ethnic origin**  
Source: Brown, C. (1984), p.197.



employed in the manufacturing industries, and these have been particularly vulnerable in the recession, the effect has been proportionately greater on British Pakistani employment. The textile industry in which over 20% of British Pakistanis were working as compared with 2% of the general population was already in decline and virtually folded in the recession. All ethnic minorities are under-represented in the distributive trades which form the largest single area of employment in the total population.

The vast majority of employed British Pakistani men are still in skilled manual and semi-skilled manual work. Their employment pattern in terms of their concentration in certain industrial sectors has not changed over the years (Anwar, 1985, p.101). In this study only four of the fathers in the intensively studied families were in full employment throughout the course of the study.

Working night shifts and overtime to increase earnings to levels comparable with workers in other industries has been a particular feature of Pakistani employment:

"Night shifts and alternating shifts are, among those regularly working shifts, more commonly worked by Asian and West Indian men than white men. As found in 1974, among black workers the incidence of shift work and night work in particular, is greatest for the Pakistanis. Broadly, then, we see that the ethnic variations in working hours and shiftwork have changed little since 1974 (Brown, 1984, p.77-79)."

Twenty seven percent of British Pakistani men were working night shifts compared with nine per cent of white men. In N.W. England, where many British Pakistani men work in the textile industry, forty five per cent of shift work was done by men from the ethnic minorities ( Smith, 1976,pp.77-79). The typical daily routine of a night shift worker is shown in fig. 2.2.

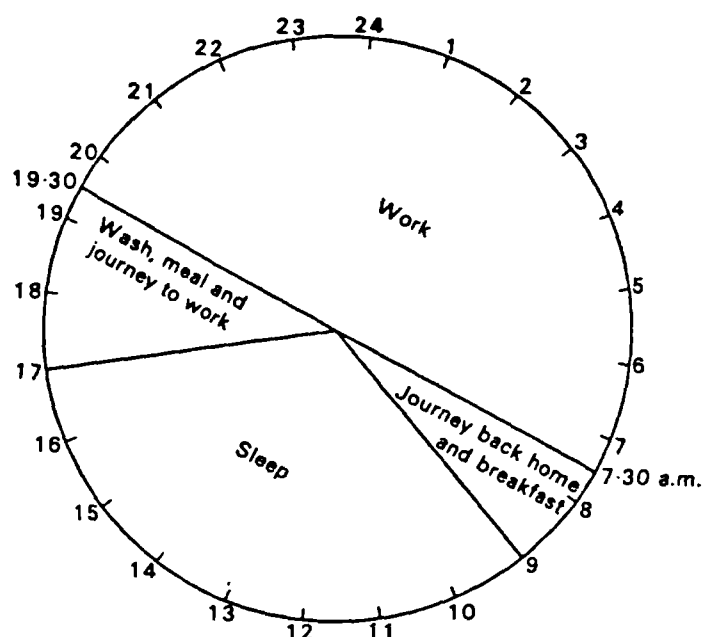


Fig. 2.5<sup>10</sup>: The typical daily routine of a night shift worker.

Night-shift work is naturally not conducive to a healthy family life; it leads to non-participation in the lives of other family members other than on rare days off. The hours are not only unsociable but also very long. Anwar (1985), in his study of British Pakistani families in Rochdale, noted that shiftwork caused health problems for the men, discord in marital relations and lack of control over growing children which greatly concerned the families.

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<sup>10</sup> Source: Anwar, M. (1985), p.122.

A growing number of British Pakistanis are self employed in places like London, Birmingham, Manchester and Glasgow as well as other cities (Anwar (1985, p.101)). In Bradford, where the majority of British Pakistanis are of rural origin, the number of Asian businesses (mainly owned by people of Pakistani origin) doubled between 1978 and 1984 from 650 to over 1,200. These were possibly initiatives to combat the high level of unemployment by pooling resources with family or kin.

Amongst professionals there are over 4,000 doctors and about 2,000 scientists of Pakistani origin working in Britain. In most cases their reasons for, and route of migration differed from that described for the majority. It is, however, not rare to find qualified teachers and graduates working in mills, as their Pakistani qualifications are not recognised in the U.K.. Of the four graduate fathers in this study three were or had been in unskilled work.

Unemployment is high among British Pakistani school leavers as with school leavers of other ethnic minorities. In 1982 while 25% of young people aged 16-19 were unemployed, the figure for ethnic minorities (including British Pakistanis) was 50% ("Social Trends", 1982). The PSI survey of 1984 confirms the findings (p.151). Discrimination against ethnic minorities in recruitment and promotion in employment is well documented in the Race Relations Act 1976, the White Paper,

"Racial Discrimination, 1975" and the PEP reports of 1975 and 1976 amongst others:

"It is fair to say that coloured immigrants were often employed in one type of job as regards low remuneration, level of skill and interest, dirtiness and heaviness, and hours of work and type of shift, and where this was true it was the most menial unattractive type of job, for which it had been impossible to attract white labour (Smith, 1976).

The PSI survey 1984 (p.15), reiterated the continued existence of the practice.

" ... insofar as these groups are confined to particular levels of jobs within specific sections of the industrial structure their chances of, for instance, escaping from certain residential areas or of their children gaining access to selective forms of secondary or further education will be minimized. They may then be incorporated more or less permanently into a position of multiple disadvantage (Anwar, 1985, p.103, referring to Allen et al, 1977)."

### Housing

Like the rest of the Britain's black population the majority of British Pakistanis live in the conurbations, particularly in the inner city areas where property tends to be cheaper and less desirable (Brown, 1984, p 76). 80% of British Pakistanis own their own homes (Brown, 1984, p.96) and tend to live in terraced property rather than any other type of dwelling (table 2.6). Following the trend of previous migrants with greater prosperity some people have been able to move to the smarter suburbs.



The 1984 PSI survey shows that on average the amount of space per person available in households of Asian origin is half that available to white households. Although British Pakistanis are not more likely to be sharing dwellings than other households of Asian origin, they do have a higher number of persons per room despite the fact that they tend to have larger properties. This is because, like the British Bangladeshis, they have large households.

The amenities available to households as part of their dwelling structure or as durable additions are shown in table 2.7. Sixty-six per cent of British Pakistanis do not have central heating in their homes, 61% do not have a washing machine and 34% have no telephone. Only people of Bangladeshi origin have less amenities than British Pakistanis.

	Column percentages						
	West White	Indian	Asian	Indian	Paki- stani	Bangla- deshi	African Asian
Lack exclusive use of bath, hot water or inside WC	5	5	7	5	7	18	5
No garden	11	32	21	15	21	56	18
No central heating	43	38	44	37	66	56	27
No refrigerator	6	6	11	4	19	37	5
No washing machine	22	37	44	38	61	78	22
No telephone	24	24	24	18	34	44	14

Table 2.7<sup>12</sup>: Amenities by ethnic group.

<sup>12</sup> Source: Brown (1984), p.109.

The economic situation and prospects of British Pakistanis is an indicator of economic and social pressures in the population and within the families. If present employment and housing trends continue the next generation of parents will be subject to similar pressures to those in the present generation. When considering non-utilisation of services it is extremely important to take into account the constraints of their work situation on parents and other family members and the fact that for families with children suffering from chronic disease the combined pressures can make life appear intolerable and impending problems insurmountable. In this context, practical help and speedy processing of claims for state benefits families can claim, for example, travel expenses and Social Services' benefits, become increasingly important in alleviating some of the pressures.

## CONCLUSION

The distinctly Pakistani way of life is quite apparent in Britain. Family units were only established in Britain when the migrants felt confident that the cultural and religious traditions embodied in that lifestyle could, in principle, be adhered to. All British cities with a sizeable proportion of British Pakistanis have several mosques, voluntary teaching of the Qur'an and Urdu, Pakistani shops and the celebration of religious and cultural events. It is a common sight to see children of school age going for Qur'an classes a couple of hours after their school day is over. The mosques are usually full for the Friday prayers and women organise their own religious and cultural gatherings in the home. Much of social life is centred around visiting between families and participation in the major events of one another's lives. Visits are made between cities to visit family, kin and biradheri, particularly at religious festivals, for weddings and during times of bereavement.

The age group of men and women in their late twenties and early thirties who are now reproducing are either the younger batch of first generation migrants or the adult children of first generation migrants. In the majority of cases both have had a strong exposure to the cultural, religious and social values of the Pakistani way of life, though for the latter who have grown up in Britain, that socialisation has been with institutions and practices which are essentially



Pakistani and muslim in nature but have been modified in Britain. Of course, this socialisation in the home and the British Pakistani milieu takes place within the context of life in Britain.

In health service delivery it is imperative to be aware of the cognitive maps and social organisation of the people the services are devised for. Careful study can illuminate features which can aid in the development and delivery of an effective and more appropriate service. This research has shown that kinship networks among British Pakistanis are channels of friendship, support and control and also channels for information. As inherited diseases are familial in nature, these channels can be utilised very effectively for the transmission of information and support between families who have children suffering from genetic diseases. This is particularly pertinent to British Pakistani families where this research shows that little stigma seems to be attached to inherited disease, and families with thalassaemic children are willing to discuss their problems and dilemmas with other families facing the same problem.

The housing and employment situation of British Pakistanis shows their position of social and economic disadvantage. When devising services it is imperative to consider this background and its implications for what a population's needs

may be and the constraints operating in the uptake of services.

PART THREE: FAMILY EXPERIENCE OF THALASSAEMIA

### THE FAMILY STUDIES

Prior to the study many health professionals attributed the low uptake rate of prenatal diagnosis for thalassaemia among British Pakistanis (table 3.1), and their attitude to the disease, to the cultural and religious dynamics within British Pakistani families and society. The typical statements made by health professionals have been stated earlier (p.16). The problems are clearly seen to lie with the families, who are perceived to be "different", and the onus for change is placed on their shoulders.

Detailed family studies revealed a far more complex situation, wherein a number of factors influenced British Pakistani families' experience of the disease and the decisions parents have made. The following chapter aims to explore these factors through the presentation of case studies which illustrate the the real issues involved and that parents made quite logical decisions given their experiences. It also reveals deficiencies in service delivery and the impact of these deficiencies on the families. It is in these areas that change is required to provide an appropriate and adequate genetic counselling service and ancillary support system. Health professionals involved in genetic counselling need to be aware of these factors when counselling families of Muslim background and pakistani origin.

% of at-risk pregnancies in which PND accepted	2nd trimester PND	1st trimester PND
20	Africans Caribbeans Pakistanis	-
40	-	Africans Caribbean
80	Indians	Pakistani
100	Cypriots	Indians Cypriots

PND = Prenatal diagnosis

People of Afro-Caribbean origin are mainly at risk of sickle cell disease. People of Indian, Pakistani and Cypriot origin are mainly at risk of thalassaemia.

Table 3.1<sup>1</sup>: Estimates of acceptability of prenatal diagnosis to counselled couples of different ethnic origins at risk of haemoglobinopathies in U.K.














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<sup>1</sup>Source: Modell et al, (1990)

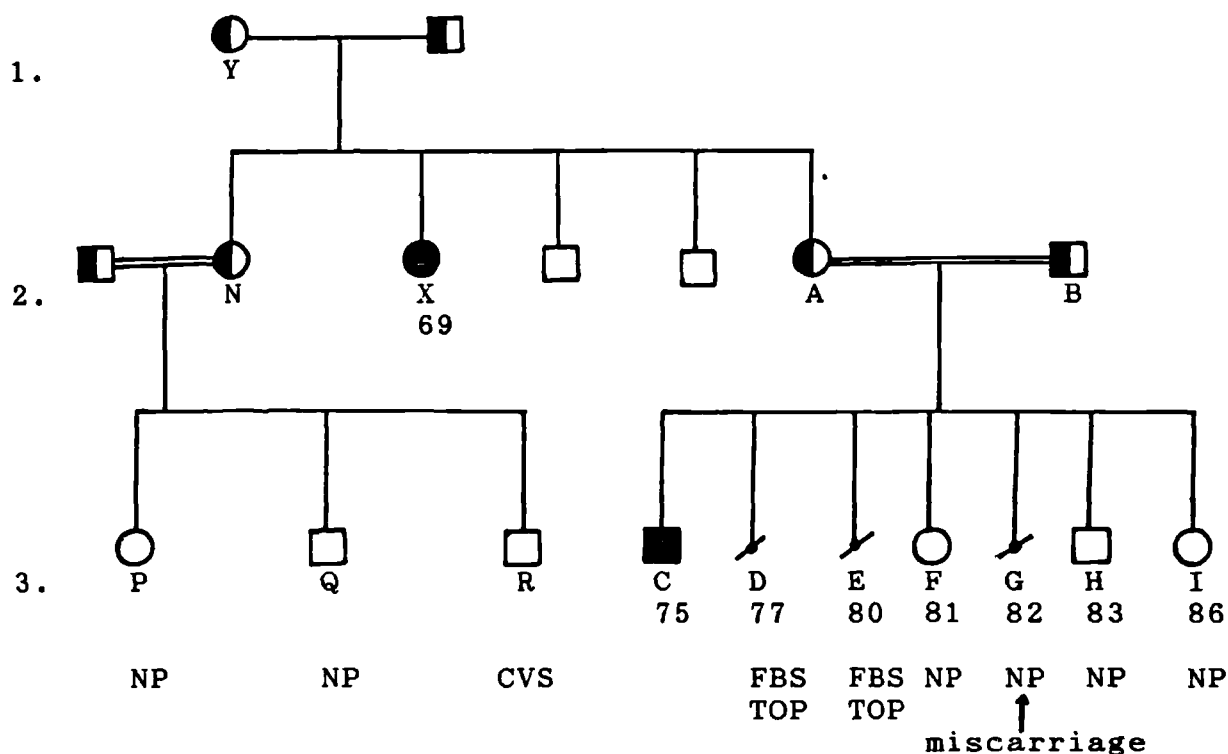
Referring to family and individual experience of inherited diseases published over three decades ago in 1955 by Reed, who introduced the term genetic counselling, Harper (1989) writes,

"Reeds case studies illustrate the background of ignorance and prejudice which his patients had to cope with and it is no wonder that he found them grateful, even when he could only give them pessimistic advice (p.3)."

Key to family trees

	Unaffected female
	Unaffected male
	Thalassaemia trait
	Thalassaemia trait
	Thalassaemia major
	Thalassaemia major
	Thalassaemia intermedia
	Thalassaemia intermedia
	Cystic fibrosis
	Cystic fibrosis
	Consanguineous marriage
	Termination of pregnancy, stillbirth or miscarriage
	Deceased
NP	No Prenatal diagnosis
FBS	Fetal blood sampling
CVS	Chorionic villus sampling
TOP	Termination of pregnancy
FC	Failure of contraception
BMT	Bone marrow transplant

The case studies were written in 1989.

Intensively studied familiesFamily O

Mrs. O. (A), born in 1951, aged 38 years, came to Britain from Pakistan in 1966 at the age of sixteen with her mother and siblings to join their father. She had matriculated in Pakistan and on arrival in England learnt English at the local college. She was then employed for several years in an electronics factory before marrying in her early twenties. In 1974 she married her first cousin (B) who had been living in Pakistan and came to Britain for the marriage. They both originate from the same city in Pakistan where Mr. O. had also graduated. The couple are fluent in English and Urdu. However they speak their mother tongue, Punjabi, at home and speak in Urdu with their children.



Mrs. O.'s youngest sibling, a sister (X), suffers from thalassaemia major. When she was diagnosed their G.P. had told Mrs. O.'s mother (Y) that her children should not marry into the family otherwise the disease would appear again. The family disregarded this information as there were many couples in the family who were first cousins and whose children were all healthy. Other than this Mrs. O. does not remember any information being given about the disease. Hence the family were unaware of its aetiology or its inherited nature.

Mrs O's first child, a son (C), was born in 1975, a year after her marriage when her thalassaemic sister was six years old. He began not to thrive from four months and showed the same symptoms as Mrs. O.'s sister, although at the time the connection was not made. From then on visits to the G.P. began. He was not very sympathetic and diagnosed the symptoms as being caused by teething. When the parents drew attention to their son's pale complexion the G.P. replied, "Well neither of you are exactly rosy-cheeked!" In the meantime the family moved to another county where they requested a blood test from their new GP. Mr. O. himself suggested that his son may have thalassaemia (at the time he knew only the name). A blood test was carried out at the hospital and C was diagnosed as having thalassaemia major at 16 months. It had taken one year to make the correct diagnosis. At the time of the diagnosis in February 1977, Mrs O was pregnant with her second child (D). Nothing was mentioned about prenatal diagnosis at the

hospital or by their G.P.. Mrs. O. learned about the disease and preventive services available at University College Hospital (U.C.H.), London, through a television programme on thalassaemia major. She contacted U.C.H. and arranged to travel to London for fetal blood sampling at 18 weeks gestation. The fetus was found to be affected. A termination was carried out in her home town by the invasive and now outdated method of hysterotomy. She was on the waiting list for a few days, the fetus was removed by a caesarian section under general anaesthetic. She then had to remain in hospital for a week - a distressing experience, both emotionally and physically. On a follow-up visit to the outpatients clinic a doctor also told her that all her children would suffer from the same condition, which made her very angry as she knew this to be false information. She said "I nearly hit him with my handbag. I didn't. Instead I got up and left and swore never to go back to him."

In 1980, having moved back to the city of their original residence where Mrs. O.'s family also live, Mrs. O. became pregnant for the third time. She travelled to London for the second time for fetal blood sampling and returned the same day. The fetus was again affected and termination of pregnancy carried out in her home town by prostaglandin. Although this was conducted in a sympathetic atmosphere it nevertheless took three days because of the danger of the rupture of the uterus caused by the scar from the unnecessary

hysterotomy. Mr. O. had phoned B.M. in despair after forty eight hours.

After the two terminations the couple desperately wanted a normal child. During a conversation with Mr. O. at this time he appeared emotionally saturated with the hospital connection and the suffering his wife had gone through. They felt they had tried everything the secular world could offer and that maybe they were not having enough faith in God to grant them a healthy child without any intervention. They had also heard of a misdiagnosis that had been made by U.C.H. and hence felt justified in not accepting all that modern medicine decreed. Mrs. O. said that morally she had never felt comfortable about terminating her pregnancies.

For the fourth pregnancy in 1981, the couple made a conscious decision not to test the fetus. Mrs. O. had also visited her family pir in Pakistan who had given her prayers to recite and an amulet to wear throughout the pregnancy. The pregnancy resulted in a healthy child (F), but they had to wait over a year before they knew from the child's condition that it was healthy, as the couple had refused neonatal diagnosis because the results were not always conclusive. They felt it better to "take each day at a time". Not knowing whether their child was healthy was very stressful as there was fear each time the child had a minor illness. However, they felt the healthy birth had strengthened their faith in God. Mrs. O.'s next

pregnancy in 1982, which she had not intended to have tested, ended in a miscarriage. Pregnant for the sixth time in 1983, Mrs. O. went to Pakistan again to visit the family pir and once again delivered a healthy child (H). By the time of this pregnancy prenatal diagnosis was available at 9 weeks using chorionic villus sampling. Mrs. O. spoke of how she had been tempted to have the baby tested at nine weeks but after conferring with her husband decided to stand by their resolve and trust in God alone. Mrs. O. was pregnant again in 1986, did not request prenatal diagnosis and delivered a healthy child (I). Seven pregnancies have resulted in one thalassaemic child and three healthy children.

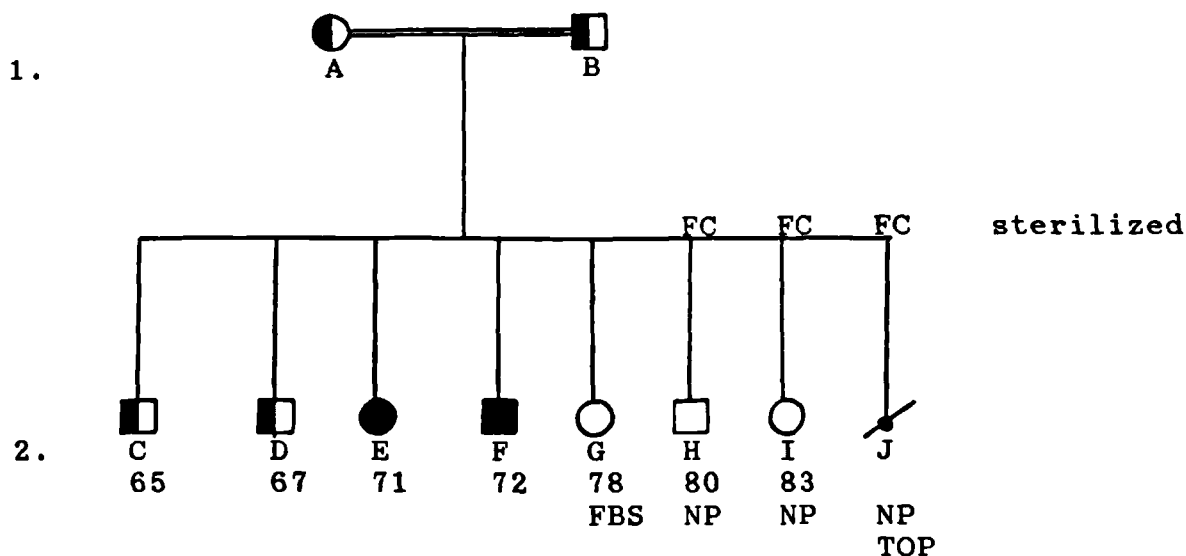
Both husband and wife have always felt termination of pregnancy to be wrong for religious reasons. Even when they had decided on a termination they felt they were committing a wrong deed but, at the time they felt they had no choice in the matter. It felt wrong to terminate a pregnancy but it also felt wrong to bring a suffering thalassaemic child into the world. They now feel they should have had enough faith in God and asked Him as their Ultimate Provider to grant them a healthy child, rather than limiting themselves to believing that their fate is determined solely by the intervention of human beings (doctors) and modern technology with its limitations. The emotional burden of coping with a thalassaemic child, the difficult terminations, misinformation, the consequent mistrust of doctors and despair

at not conceiving a healthy child also prompted them to seek an alternative course of action. The couple have also been concerned about the likelihood of misdiagnosis and the inability of the tests to distinguish between thalassaemia major and thalassaemia intermedia which they believe to be milder in its manifestation.

The experience of this couple had an important impact on the attitudes of other at-risk family members. Mrs. O.'s married younger sister (N) is a qualified teacher. During much of the research period she had politely but firmly declined any discussion of her own situation. On one occasion when the researcher was visiting Mrs. O.'s mother N was also present. She told the researcher not to discuss their thalassaemic sister's condition with her mother as she was a widow and already worried about her. The effect on family life of her sister's condition plus the experience of Mrs. O. had clearly created a barrier to discussions regarding thalassaemia and surrounding issues (denial). She appeared to have delayed an assessment of her own situation until she could cope.

N became pregnant only after Mrs.O. had visited the family pir and delivered a healthy baby without undergoing prenatal diagnosis. N also visited the same pir before embarking on her first pregnancy. She too delivered a healthy child (P). Subsequently N has had another healthy child (Q) without C.V.S.. When pregnant for the third time she requested C.V.S.

as she felt she could no longer cope with the anxiety during pregnancy. The child (R) is healthy. However, N says she would have terminated the fetus if it had been affected. This case study illustrates not only that attitudes towards a service can change but that they also oscillate over a period of time, both with developments in technology and with people's life experience. The case also shows that formal education does not necessarily denote an acceptance of modern technological developments, as is often supposed. It also shows that medicine is self-defeating when it is associated with insensitivity, misinformation and mismanagement.

Family G

Mr. G. (B) is fifty two. He was born in Pakistan where he matriculated and qualified as a teacher. He is of an urban background, is literate in Urdu and has a working knowledge of English. His wife (A), aged forty three, is his first cousin and was born in the same city as her husband in the Punjab where she studied to the eighth grade. She is also literate in Urdu and has a working knowledge of English. The family speak Punjabi at home. The couple were married in 1964, in Pakistan, and immediately migrated to Britain where Mr. G. found employment in a factory.

All seven children in this family were born in Britain. The couple had no knowledge of thalassaemia major until the birth of their third child (E) in 1971. She was normal at birth but by three months was not thriving. The couple visited their G.P. who prescribed different medicines at each visit.

Mrs. G. found the G.P.'s attitude very casual as she felt he did not take any notice of her worries. The child was eventually referred to the hospital by the G.P. at six months where she was diagnosed as having thalassaemia major. Mrs. G. does not remember their having been told anything about the disease except that their child was to receive regular six weekly transfusions.

At the time of the diagnosis Mrs. G. was already pregnant with her fourth child (F) who also suffers from thalassaemia major. These births occurred in 1971 and 1972 respectively when prenatal diagnosis was not yet available. For the next six years Mrs. G. avoided pregnancy by using an I.U.D., for fear of having another thalassaemic child. During this period she does not remember having been given any further information about the disease.

Mrs. G. says she had wanted more healthy children and hence after six years became pregnant in 1978 hoping the child would be healthy, as she had previously conceived two healthy children. During her pregnancy she remembers the paediatrician suggesting that she should terminate the pregnancy as all her future offspring would suffer from thalassaemia and that the condition was caused by the fact that she and her husband were first cousins. Prenatal diagnosis was suggested and Mrs. G. remembers feeling pressurised by the staff nurse to undergo the test. The



staff nurse also remarked that she could not understand why Mrs. G. wanted so many children.

Mrs. G. agreed to prenatal diagnosis under pressure from the staff nurse and hospital doctor, to put an end to the discussion though she knew inwardly that on no account was she going to terminate the pregnancy. The fetus was diagnosed to be affected and Mrs. G. decided against termination. The staff nurse rebuked her, saying she had been highly irresponsible by going through with the test and had wasted £2000 of public funds.

The couple are against termination of pregnancy for religious reasons, unless the mother's life is in danger. Mrs. G. says she felt that if she was fated to have another thalassaemic child, then she would look after it as she was caring for the others. She felt that the medical professionals' insistence on terminating an affected fetus demeaned the lives of her two thalassaemic children. She said, "My children require only blood to function, otherwise there is nothing wrong with them. There are other handicapped children much less fortunate than them and they manage to live." At the time neither of the thalassaemic children were receiving Desferal, nor was the prognosis clear to the parents.

Mrs. G. remembers this pregnancy as being agonising, knowing she was carrying an affected child. She visited a pir in the

city where she lives for an amulet which she wore throughout her pregnancy and constantly prayed for God's blessing. It transpired that the fetus had been misdiagnosed and Mrs. G. had a healthy child (G).

Mrs. G. had decided at this point not to have any more children because of her distress during the pregnancy. The couple were unable to use anything but the condom as contraception as Mrs. G. was taking medication for a clotting tendency associated with the delivery. However, seventeen months after the previous birth Mrs. G. became pregnant in 1980. She did not inform her G.P. or the hospital about the pregnancy for six months because of the pressure she felt she had been put under during her previous pregnancy. She had found the hospital doctor to be forceful and insensitive in his manner. Hence, before visiting the hospital again she telephoned the receptionist to ensure that she would not have the same doctor. She gave birth to a healthy child (H) in 1980.

At this point she was seriously considering sterilization but felt afraid of the side effects she had heard about from other women who had undergone the procedure. Failure of contraception for the second time resulted in yet another pregnancy two years later. Again she did not inform the G.P. or hospital until the last few months of the pregnancy, and gave birth to another healthy child (I). Prior to the

delivery she had signed the forms requesting sterilisation but again suffered from venous thrombosis and the sterilisation was not carried out. She was to be on medication for one year during which time she had the same problems with contraception and conceived within that period. Mrs. G. chose to terminate this pregnancy (J) at nine weeks. She was aware that medically it was not safe for her to go through another delivery. Sterilization was carried out after the termination.

Mrs. G. says she had wanted a large family. Having had two healthy children and two thalassaemics the couple decided to have one more child. Due to the distress she experienced during this pregnancy she decided not to have any more children. Her three subsequent pregnancies were not planned but resulted from a combination of her medical condition, consequent postponement of sterilization and failure of the sole contraceptive method the couple were able to use. Through the last three pregnancies carried to full term she has visited a pir for amulets and prayers and sought support from her faith to cope emotionally.

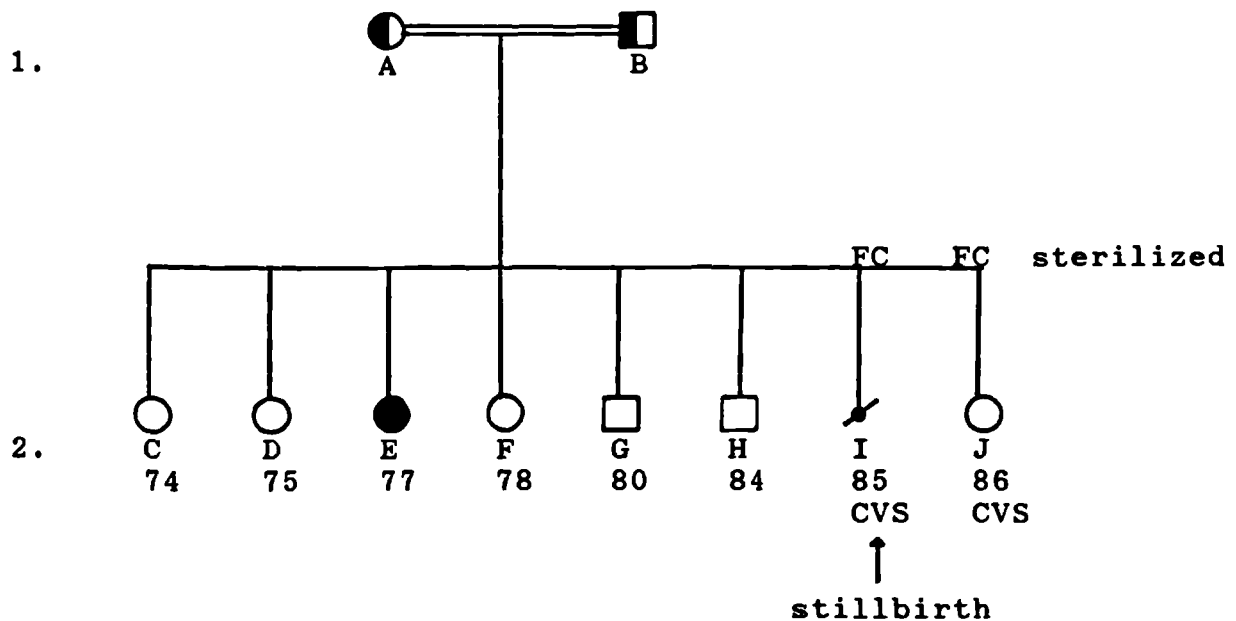
The couple both adhere to the belief that it is wrong to terminate a pregnancy unless the mother's life is in danger. However, now that the thalassaemic children are in their teenage years, on regular Desferal therapy, their growth is stunted and their sexual development delayed Mrs. G. fears for

their future, whether they will be able to marry and lead healthy sexual lives. She is not confident that she will be able to arrange marriages for her thalassaemic children given their medical condition. With hindsight and the existence of C.V.S., she feels she cannot be as emphatic about termination of an affected fetus as before and would leave her children to decide for themselves.

The couple consider it imperative to know the carrier status of their children. Their most preferred form of prevention would be to test for carrier status before marriage and to arrange marriages accordingly. However, with their eldest son (C) the couple have had to concede to pressure from the extended family in Pakistan to arrange a match with his first cousin without testing her carrier status. The extended family do not yet understand the inherited nature of the condition and its implications. The match was desired by the common grandmother of C and his fiancée and if couple G. were to refuse their niece's hand they would appear to be disconnecting themselves from their extended family. The issue was resolved by couple G. and C deciding that his fiancée will be tested immediately after the marriage. Mrs. G. asked me to arrange blood tests for her brother's children who live nearby so that she would know their carrier status and hence be able to ask for the hand of an unaffected niece for her other carrier son (D). She felt the suggestion to test for carrier status in the extended family would be better

coming from an outsider, who could also explain the relevance of the condition for the whole family.

The couple, as well as the children, have used every opportunity to gain more knowledge about thalassaemia and have been regular attenders at the thalassaemia support group meetings.

Family T

Mrs. T. (A), born in 1955, aged 34 years, has lived in Britain for fifteen years, whilst her husband arrived 25 years ago at the age of ten. They are double first cousins<sup>1</sup>, both originally from a rural background. Mrs T. went to school for three years as a child and is literate in Urdu. She understands a little English. Mr. T. is literate in English, speaks Punjabi fluently and is able to read a little Urdu. He arrived in Britain with his father at the age of ten and remained at school until the age of sixteen. He then began work at a textile mill and returned to Pakistan in 1971 to marry at the age of seventeen. He now works in a factory doing shift work.

All seven children in this family were born in England. The

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<sup>1</sup> See p. 267

couple's third child (E), born in 1977, has thalassaemia major. The child was unwell from birth and discharged from hospital after five weeks, undiagnosed. At five months she still weighed only ten pounds. The staff at the clinic advised Mrs T. to take her daughter to her G.P. who arranged for a blood test and thalassaemia major was diagnosed. The first blood transfusion was given when E was six months. Mrs. T. does not remember having been told anything about the disease nor does she remember any time that an interpreter was used at the hospital. She had thought her daughter would require only a few transfusions. During her next pregnancy she remembers the paediatrician commenting that she could have another thalassaemic child. She also remembers him telling her that the condition was caused by herself and her husband being cousins.

The following year, in 1978, Mrs. T. gave birth to their fourth daughter (F), who is healthy. At this time, during a hospital visit, Mr. T. was told that treatment for thalassaemia meant life long blood transfusions and that future offspring were also at risk. He decided that he did not want any more children. However, Mrs. T., having given birth to four girls, wanted to have a boy also. Her desire was linked to concern for her own and her husband's old age, when the social responsibility of caring for parents lies with sons rather than daughters, who generally marry and move away to live with their husbands. She was also worried about her

own marriage. Her concern about the latter was influenced by the fact that in Pakistan a close male relative had been pressurised by family members to take a second wife, so that he could have a son in the family. Also, her own mother had not borne any sons and had been made to feel deficient. Mrs. T. herself commented that it is very difficult to escape the fears of one's conditioning. Despite her husband's assurance that this would not happen in her case, she nevertheless desired a son for her own marital security. Mr. T. was using contraception but pregnancy occurred, the couple being in Pakistan at the time. Although they do not have a pir either in Pakistan or England Mrs. T. did visit a shrine to pray. During the pregnancy she says she felt afraid and prayed constantly for a healthy child. In 1980 she gave birth to a healthy boy (G).

For the next three years Mrs. T. used an I.U.D.. During this period she says she felt she desperately wanted to have another child in the hope that it would be a boy and a companion for her son, even though her husband did not want any more children. She said that although she was aware of the risk, four of her five children were healthy. She became pregnant immediately and gave birth to another healthy son (H). Mrs. T. had the I.U.D. inserted again after the birth and did not intend having any more children. However, despite the I.U.D. she became pregnant again after thirteen months in 1985. The I.U.D. was left in her womb for fear of disturbing



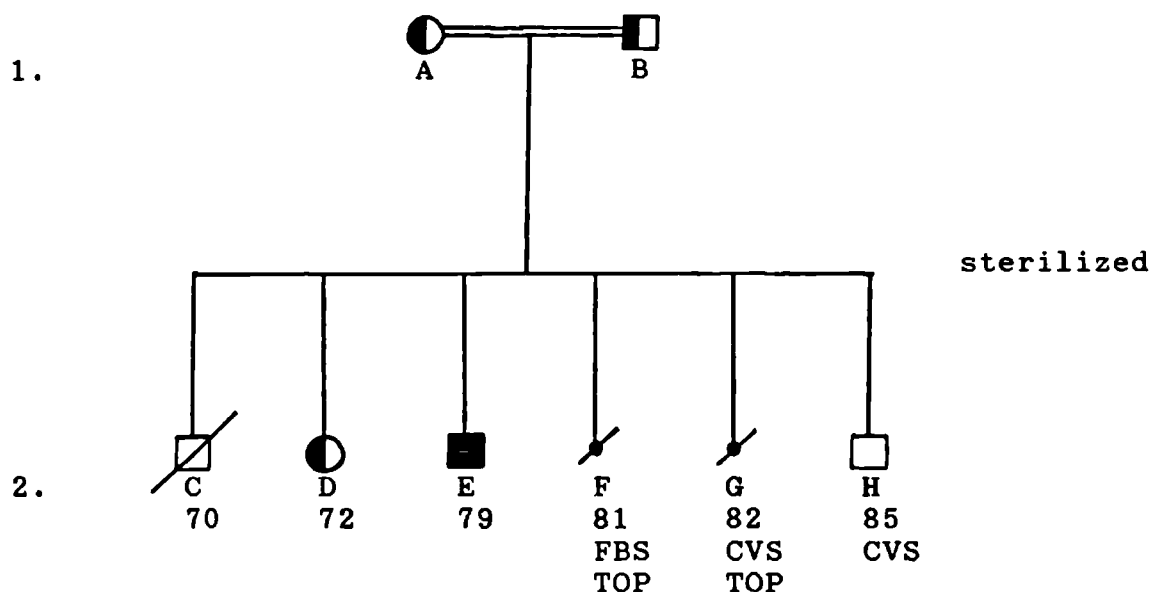
the pregnancy. Pressurised by her husband she made an appointment to terminate the pregnancy but as the date approached she cancelled the appointment because she felt she could not go through with it. She believed it to be wrong to terminate a pregnancy, for religious reasons. The hospital doctor who deals with this family had noticed from the case studies that Mrs. T. had booked for a termination of pregnancy and then cancelled it. A home visit was made where Mr. T. was clearly distressed about the pregnancy and on being informed about prenatal diagnosis wanted the fetus to be tested. Mrs. T. did not want to terminate the pregnancy. On being told that she and her husband could make their final decision after the test, she consented. She was also reminded that there was a 75% chance that the child would be healthy which would remove the couple's anxiety. Her negative reaction to prenatal diagnosis was based on knowledge of Mrs. G.'s experience (previous case study) wherein Mrs. G. had felt pressurised to agree to a test and a termination. Mrs. T. was under the impression that if one had the test then termination of an affected fetus was compulsory. Mr. T. had not been aware of the availability of the test, for as he did shift work, he had not attended any of the thalassaemia support group meetings where it had been discussed.

The test was to take place in a nearby city, I escorted Mrs. T. to the hospital for moral support. Chorionic villus sampling was carried out transabdominally in a supportive

atmosphere. The fetus was found to be unaffected. However, the child (I) was born dead at seven months gestation. Mrs. T. requested sterilisation but was told to wait three months by the doctor as she was bereaved. During the three months Mrs. T. became pregnant yet again. She requested C.V.S. and delivered a healthy child (J) and asked to be sterilised immediately afterwards. She was relieved at not having to face the dilemma of whether or not to abort an affected fetus.

On first learning about the treatment for thalassaemia major and the genetic risk to future offspring Mr. T. had decided he did not want any more children. For the reasons outlined above Mrs. T. still desired male offspring. She remained against termination of pregnancy for religious reasons but was relieved to be able to utilise prenatal diagnosis to find out if the child was affected or not. The couple were able to keep the situation private as the hospital visits did not require an overnight stay. The fact that the test was performed transabdominally rather than transcervically probably made it more acceptable. Mrs. T. says, however, that she feels it is entirely dependant on a couple's situation and beliefs as to what they choose to do regarding prenatal diagnosis. She added that it was important to know the carrier status of each child from birth. She felt that it was most probable that in marrying, people would marry first and then be tested.

Family P



Mr. and Mrs. P. are of an urban, educated background. Mr. P. (B), aged forty eight years, born in 1941 in Pakistan, holds a prominent position within a minority Muslim sect in the city in which he lives. He is a graduate and spent a further seven years studying theology in Pakistan. He has also lived in various countries where he was posted as a missionary. He is fluent and literate in Urdu and English. Mrs. P. (A) aged forty two, born in 1947 in the same town as her husband, is literate in Urdu and has a working knowledge of English. They speak Punjabi among themselves in the home but speak Urdu with their children. The couple married when Mr. P. was 28 and Mrs. P. 24 years old. They are first cousins.

The couple's first three children (C, D & E) were all born in Pakistan. C, born in 1970, weighed only 3lbs at birth and died at the age of two months. In retrospect the couple think

he may have been thalassaemic as he was very anaemic but this is unlikely as thalassaemia rarely presents before four to six months of age. D was born in 1972, and was also a small baby weighing only three and a half pounds. She is now known to be heterozygous for thalassaemia major. The third child, (E), was born in 1979. He was well at birth but contracted a chest infection at one and a half months. Mrs. P. had been staying with her parents in Karachi, a major city in Pakistan, when this occurred. E remained unwell despite medication. On the suggestion of a paediatrician who suspected anaemia he was given a blood transfusion at the hospital. Blood tests were done but no diagnosis offered. E became well after the transfusion and Mrs. P. went back to her home in the Punjab where E became weak again. A doctor prescribed some medicine but also suggested that E have a blood test. For this the couple travelled to Lahore, a large city fifty miles away. Thalassaemia major was diagnosed and the couple told that E would require regular blood transfusions for a lifetime. E was three months old at the time. Mention was also made of bone marrow transplantation for which the doctor said E was, as yet, far too young. Desferal was not mentioned.

The child was started on regular blood transfusion, and from the beginning the couple were very careful about where the blood for transfusions was obtained. Initially, relatives gave blood, but then Mrs. P. found it difficult to make repeated requests. Blood was then obtained from the Naval

Hospital where the charges were 250 rupees (approx. £25) per unit (about one pint). The couple's income at the time was 500 rupees per month.

As her husband had been posted overseas Mrs. P. moved to Karachi to stay with her father. Blood was still expensive. She had also been told that E needed Desferal, which she was able to purchase only a few times due to its scarcity and expense. Sometimes the cost of the treatment amounted to over two thirds of her income. A doctor from the hospital appealed in the press for healthy people to donate blood for E and the plea was successful. Help was also offered in the form of blood from a visitor to the hospital who had asked about Mrs. P.'s problem. Mrs. P. says of that period that although it was extremely difficult she found that as soon as one door closed God would open another. Mr. P. was posted to a city in England in 1980. His wife and two children joined him in 1981.

My first contact with the family was when Mr. P. attended the initial introductory meeting at the hospital in September 1981 where he informed us that Mrs. P. was two months pregnant. The couple were not aware of the inherited nature of the condition or of the availability of prenatal diagnosis. When asked whether they wanted any more children he immediately replied, "Not like this." He asked us to his home to discuss prenatal diagnosis further. The couple decided

immediately after the discussion that they wanted the fetus to be tested and would request a termination if it was found to be affected. The couple adhere to the belief that termination of pregnancy is permissible in Islam only if the mother's life is in danger or for medical reasons. They felt the burden of the disease on the child and family was such that termination of pregnancy was justified. It is worth noting that the difficult experience of coping with the disease in an economically developing country was probably a factor affecting the attitude of this couple to the disease and the lack of hesitation in making their decision.

Fetoscopy was carried out in London at 18 weeks, and the result was positive for thalassaemia major. The termination was carried out in the city of residence and required a hospital stay of three days. At a follow up meeting with the couple in their home, Mrs. P. appeared composed as she talked about the termination. At the next visit when Mrs. P. and I were alone she cried. She had found the experience distressing as the child had been fully formed. She was willing to undergo the same test only once more in an attempt to conceive a healthy child. She was also upset as she had only just internalised the fact that the condition was hereditary and that this had implications for every member of the family. She had recently discovered that her sister and future brother-in-law (Mr. P.'s brother) were both carriers. They had been prompted to find out their carrier status

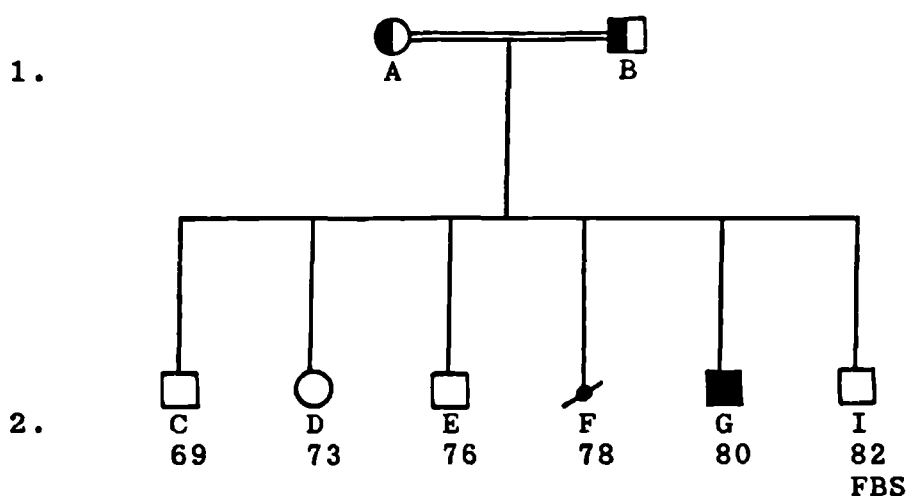
through correspondence with Mr. and Mrs. P.. This couple, however, had decided that they would rather not have children than not marry one another.

Couple P. have attended the thalassaemia society meetings regularly where they were informed that prenatal diagnosis would soon be available at 9 weeks by chorionic villus sampling. They decided to plan the next pregnancy after the test became available and were the second couple to use the test. In 1982 they travelled to London. After a discussion in which the very early stage of development of the test was explained and understood, chorionic villus sampling was carried out. The fetus was affected and the pregnancy terminated in London because of the need to confirm the diagnosis by taking blood to verify the new D.N.A. method. Whereas Mrs. P. had been very upset after the earlier mid-trimester termination, she was much calmer after this termination. She said there was a world of difference in the two experiences. First trimester testing meant that a strong emotional bond had not been established with the fetus and it also allowed privacy and discretion. Had this been a mid-trimester termination she would not have thought of conceiving again. The couple planned a pregnancy in 1984, requested first trimester prenatal diagnosis and a healthy child (F) was delivered. Mrs. P. underwent sterilisation after the delivery.

The issue of the medical implications of consanguineous marriage had been raised by Mrs. P. herself during one of the home visits. Doctors in Pakistan and in England had implied that thalassaemia was caused by consanguineous marriage. Her own feelings were that this was ridiculous as there were many consanguineous marriages in her own family but this was the first incidence of disease or handicap. She had also questioned women in the hospital in Pakistan with handicapped or thalassaemic children as to whether they were related to their husbands or not; some said they were and some said they were not. However, after discussion the couple did understand the genetic implications of consanguinity, the need to be aware of carrier status and the options available.

The couple felt it was important to know the carrier status of the children at birth. They thought being informed at school-age or just prior to marriage would be too difficult emotionally. Mr. P. stated that both he and his wife would be happy to discuss their experience with other families but he wished to be approached as a parent and not on account of his prominent religious position. The decision regarding termination of pregnancy had been a personal one and other people had to make their decisions based on their own beliefs and situations. Many members of the couple's extended family in Pakistan and in the United States have been H.L.A. typed as possible donors for a bone marrow transplant for E but, as yet, a compatible donor has not been found.



Family R

Mr. R. (B), born in 1934 is fifty four. He came to Britain in 1962 at the age of twenty eight. He is of rural background and had five years' schooling in Pakistan. He is literate in Urdu but speaks hardly any English. In 1968 he returned to Pakistan and married his first cousin once removed<sup>1</sup>. Mrs. R. (A), born in 1952 is thirty seven and was born in the same village as her husband. She has not been formally educated and speaks only Punjabi. Their first three children (C,D & E), born in 1969, 1973 and 1976 are all healthy. The fourth child (F) died at the age of six months. The fifth child (G), born in 1980 in Pakistan, has thalassaemia major. He was born during a period when the couple had returned to Pakistan to look after Mr. R.'s father who was ill. The child was well at birth but after he was circumcised at one month his health deteriorated. The family initially thought that the circumcision was the cause of his ill health. He was taken to

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<sup>1</sup> See p.267

a number of doctors and hakims<sup>2</sup> who prescribed various medicines but the condition remained undiagnosed. Visits were also made to shrines for blessings. Paediatricians were consulted, for which the family had to travel from the village to the nearest town from the village. G was diagnosed as suffering from thalassaemia at eight months and given his first transfusion. Initially family members gave blood but then it had to be bought. Again, they had to travel to the nearest town for the transfusion. Often the cost of the travel, consultation, transfusion would be about 500 rupees. The family's monthly income at the time was 1000 rupees.

The couple decided they had no choice but to return to England, which they did in 1982. They did not know anything about the condition except that their child required blood transfusions for a lifetime. When I first met the family G was receiving blood transfusions and Desferal. Mrs. R. was pregnant but the family did not know that the condition was hereditary. They were informed of the availability of prenatal diagnosis, at that time available only at eighteen weeks. They expressed an interest in the test and wished to meet another couple who had utilised the test, and this was arranged. They decided to go ahead with the test, which was carried out in London. The fetus was unaffected and the pregnancy continued. Had the fetus been affected the couple would have terminated the pregnancy. Since the delivery

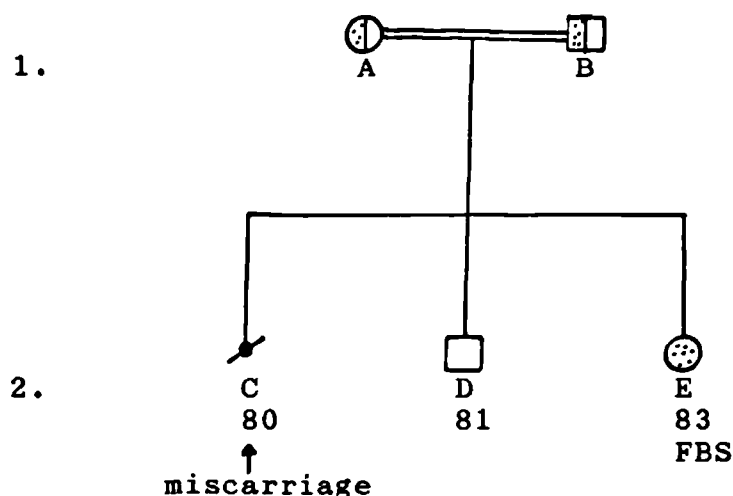
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<sup>2</sup> Practitioners of South Asian indigenous medicine.

Mrs. R. has had an I.U.D. inserted and the couple do not want to have any more children. Their reason for not wanting sterilization is that they may be left childless if all their children were involved in an accident.

Mr. R. has attended the thalassaemia support group meetings regularly, sometimes with other members of the extended family. This has entailed a journey of forty miles each time. Bone marrow transplantation was discussed at one of the meetings and since learning of the operation as a possibility of a complete cure Mr. R. has tried every avenue to find a compatible donor. He has had all his family members tested and has himself arranged for possible donors from the family in the village in Pakistan to be tested and for blood samples to be flown to England. Unfortunately a compatible donor has not yet been found.

The couple would like to settle in Pakistan so that they can look after the elderly father but feel unable to do so because of their child's condition.

Family I

Couple I. are of rural background from Pakistan. They are both literate in Urdu but do not converse well in English. Mr. I. (B) is aged thirty three and was born in a village in Pakistan in 1956. He has lived with his parents and siblings in England since 1969. He used to work in a textile mill but since 1983 has been unemployed. His wife, Mrs. I. (A) is his first cousin once removed. She is thirty and was born in 1959 in the same village as her husband. They were married in 1980 in Pakistan and then the couple returned to England together. The couple live in a large terraced house with Mr. I.'s sister, her husband and their child. Mr. I.'s parents and other siblings live in the same street. The family also carries the gene for cystic fibrosis and one of Mr. I.'s sisters suffers from the disease.

Mrs. I.'s first pregnancy, in 1980, ended in miscarriage. Pregnant again in 1981, she was prospectively diagnosed as

being a carrier at the ante natal clinic. It was not possible to test her husband at the time as he was in Pakistan. When he returned and was found to be a carrier it was too late for prenatal diagnosis. The child (D) is normal.

The initial visit to the couple's home was with the haematologist who had notified me of the case. Mrs. I's mother-in-law and sister-in-law were also present when the visit was made and joined in the discussion. The nature of the disease, the pattern of inheritance and the facilities available for prevention were explained. The family were not aware that cystic fibrosis was inherited in the same manner, so the explanation was applied to that also. Further visits were also made to the home when Mr. I. was also present. Blood tests were carried out at this stage which showed that prenatal diagnosis could be done by chorionic villus sampling in the future.

The couple had learned of prenatal diagnosis from me, so when doctors mentioned it later at the hospital they were confident about the facts. Prenatal diagnosis had also been discussed at the thalassaemia meetings and Mrs. I. had heard other women relating their experiences.

On learning that she was pregnant, in 1983, Mrs. I. immediately informed her G.P. and asked for arrangements to be made for prenatal diagnosis. However he told her that no such

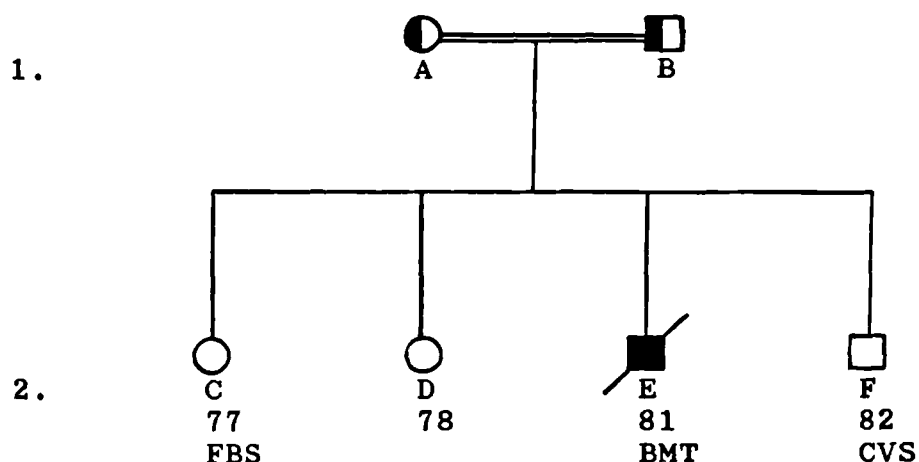
test existed. She was not able to contact me as she knew me to be overseas at the time. Mrs. I. was routinely referred to the hospital by her G.P. at sixteen weeks, and by this time the pregnancy was over the limit for chorionic villus sampling, and she was referred for fetal blood sampling at eighteen weeks. Mrs. I. travelled to London, accompanied by her sister-in-law, for the prenatal diagnosis. As Mr. I. was unemployed it was necessary to obtain a travel warrant covering the travel expenses from the Social Services Department. Speedy arrangements had to be made.

The fetus was found to be affected. By the time the couple knew of the results Mrs. I. was 20 weeks pregnant. When the couple were deciding whether or not to terminate the pregnancy they visited another couple with a thalassaemic child who had experience of prenatal diagnosis. The issue was also discussed with Mr. I's parents. The couple decided not to terminate the pregnancy, as it was too advanced. Mrs. I. said that the pregnancy was apparent and the neighbours as well as the family knew of it. The couple felt very angry towards the G.P. as the pregnancy would have been terminated had it been detected in the early stages. The affected child (E) has thalassamia intermedia and not thalassaemia major. The distinction between the conditions can not yet be made through prenatal diagnosis. The child does not have transfusions, as yet, but Mrs. I. still feels the burden of the disease as the child is very small, her skull has become enlarged and there

is always the possibility that she may require regular transfusions. Even at the age of three E was withdrawn at the nursery and asked why she was smaller than the other children. The couple intend to use chorionic villus sampling to have healthy children. Mrs. I. felt that first trimester diagnosis afforded greater privacy and there was less attachment to the child. They would not consider another pregnancy if the prevention service was not available. The couple use contraception to control conception. They feel justified religiously in terminating an affected fetus as they think the disease causes too much suffering. They did not feel it necessary to consult a pir, though they do seek such advice on other matters.

The family are very open about the condition. The family in Pakistan and in England have been told about it as well as neighbours and friends who ask. The couple feel that it is a situation that other people are also confronted with, so there is nothing to hide. They would be interested in a bone marrow transplant as a complete cure.

Mrs. I. felt it was important to know one's carrier status before marriage so that marriages between heterozygotes could be avoided. She felt it was very important to know the carrier status of one's children from birth.

Family K

Mr K. (B), aged thirty four, born in 1955, is of urban background. He studied in Pakistan until the age of thirteen and then joined his father in England in 1968. He remained at school until the age of sixteen and then worked in a factory until he was made redundant in 1986. He is literate in Urdu and English. Mrs. K. (A), aged thirty three, born in 1956, originates from the same city as her husband in Pakistan and is his first cousin. They were married in Pakistan in 1974. Mr. K. returned to England two months later and Mrs. K. followed a year after. She studied to the ninth grade in Pakistan, is literate in Urdu and now has a good working knowledge of English. The couple were picked up prospectively when Mrs. K. was pregnant for the first time in 1977. They were given information about the disease and the availability of second trimester prenatal diagnosis. They were also told of the risk to every pregnancy which they understood. Mrs. K. underwent the test, but at twenty three



weeks gestation, as there had been a miscalculation of the dates. It would have been too late to terminate the pregnancy even if it had been affected. Fortunately it was not, and Mrs. K. gave birth to a healthy daughter. Mrs. K.'s parents in Pakistan had been informed of the couple's situation and her father had prayed at a shrine for the child's health.

Contraception had been discussed at the hospital and soon after the delivery Mrs. K. began to take the pill. She suffered side effects and discontinued its use. She became pregnant in 1978 and attended the ante-natal clinic at three months. No mention was made by the staff of her risk or of prenatal diagnosis. Mrs. K. says that she did not speak English at the time and although they knew of their risk all the information seemed unreal and was put to the back of their minds. The couple had never heard of thalassaemia or inherited disease before the previous pregnancy. They also did not know of any other at-risk couples. Mrs. K. gave birth to another healthy daughter (D).

Mrs. K. became pregnant again in 1981. They wanted a small family and were hoping for a boy. At four months gestation Mrs. K. went on holiday to Pakistan for two months. The couple had had two healthy children and had "forgotten" about their inherited risk. On her return she attended the ante-natal clinic where she saw a doctor as she was not gaining weight. The doctor knew she was at risk for thalassaemia but

Mrs. K. was six months pregnant and it was too late for prenatal diagnosis. E's cord blood was checked regularly after birth and was diagnosed as thalassaemic at five months. He underwent bone marrow transplantation in 1982 and died during the procedure.

After the transplantation Mrs. K. said that after seeing so much suffering all the facts relating to the condition become imprinted on the mind. Mrs. K. became pregnant again in 1982. The pregnancy was not planned. Mrs. K. was unable to use the pill and the couple had been using the sheath as contraception. They were not worried about the risk of pregnancy as they knew they would have the fetus tested. Mrs. K. underwent chorionic villus sampling and delivered a healthy son (F).

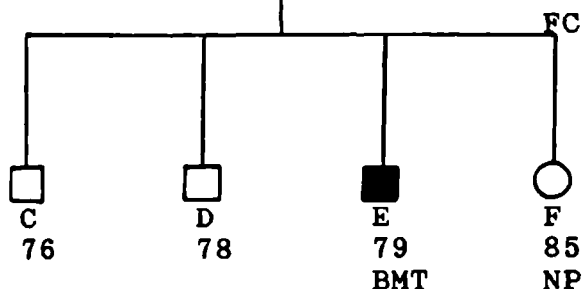
With their experience of the disease the couple felt it was best to test the fetus, especially as the test was painless. They also stated that they would have terminated a pregnancy with an affected fetus if the test had been available even at eighteen weeks.

Family Z

1.



2.



Mrs. Z. (A), aged 33, born in 1956, came to England in 1970 with her mother, three brothers and two sisters. Her father was already living and working here. The family is from a small town in the Punjab. She studied to the seventh grade in Pakistan and is literate in Urdu. She speaks only a little English and attends local English classes. Mr. Z. (B) is her first cousin. He is aged 35 years, born in 1954, came to Britain in 1965 and is from the same town as his wife. The rest of his family live in Pakistan. He graduated in Pakistan and has a working knowledge of English. The couple were married in 1975 and Mr. Z. began work in a textile factory. He was made redundant in 1981.

The couple's first two children (C and D), born in 1976 and 1978 are both healthy. The third child, born in 1979 has thalassaemia major. The child was well at birth, weighing

eight pounds. After a few months he began to cry a lot and Mrs. Z. thought the baby was suffering from stomach pains. By the time the child was one year he was drinking very little milk at each feed and was very pale. The couple made many visits to the doctor who prescribed medication but would not refer the child to the hospital. Mrs. P. says the G.P. would not take any notice of her but she knew there was something wrong with her son. She felt very frustrated at not being able to demand more attention from her G.P. due to not having a command of English. She says she spent what seemed like endless nights holding the baby and crying when she couldn't sleep. "I felt so much pain", She says, "but there was nothing I could do." Eventually, a caring nurse at the children's clinic that Mrs. Z. was visiting noticed the child was very ill and telephoned the G.P. to ask him for a hospital referral. The appointment was for three months later and the same nurse rang to have it changed to an earlier date. The child was diagnosed as suffering from thalassaemia major at fifteen months. By this time he had also developed an enlarged spleen.

Mr. Z. remembers the paediatrician explaining the condition in terms of E having "bad germs" in his blood. When he learned the real cause of the disease he was extremely annoyed as he felt he had been misinformed. At the time, I, who was informing Mr. Z. had not realised that the doctor had probably been referring to "bad genes". Mr. Z. also remembers a doctor

telling him the condition was caused by cousin marriage. At a later date he was given this explanation by a doctor in Pakistan too. The child began to have regular transfusions but the parents found it difficult to administer the Desferal at home. Mr. Z. commented that he found the constant hospital visits very time consuming and had to take time off work to take E to the hospital as his wife had to look after the other children. He felt that their lives seemed to revolve around their son's illness and this detracted from the needs of the other children. He said he was not prepared to disrupt the lives of the other children with the upheaval caused by inserting the needle at home every night as his son became anxious and cried. The couple were aware that the intramuscular injections at the hospital were not as effective as the pump. Due to their worries about Desferal Mrs. Z. had visited a mother she had met at the thalassaemia support group meetings to discuss the matter. The couple decided they would wait until their son could understand the need for the pump and then start to use it in the home.

At one home visit Mrs. Z. had kept her healthy son, D, at home for me to examine. She said that he was pale and suffered from constipation and she feared that maybe he would develop thalassaemia.

In 1981, E was given a course of steroids as it was suspected he may have thalassaemia intermedia and not thalassaemia

major. None of the other children known to the couple through the support group meetings had had this treatment. Although E became bloated whilst taking the steroids he was able to go long periods without having a transfusion. At a home visit Mrs. Z. was elated as she thought E was going to be cured. However, after the course of steroids, he still required regular transfusions. This caused confusion with the parents and they thought that maybe their son did not suffer from the same condition as the other thalassaemic children. They also felt that the doctors did not really know what they were doing, as they kept on changing the diagnosis.

Despite at times appearing to understand the cause of thalassaemia the couple have not totally accepted that their child suffers from this particular condition. They appear to sway between believing and not believing that their affected child has thalassaemia. They do not accept that inheritance has had any part to play in the occurrence of the disease. It appears that this is caused by the fact that the child has undergone treatment which has differed from the other thalassaemic children; that information given to them needs to be made clearer and that misinformation regarding the disease has made them doubtful about accepting the medical profession's word. The couple constantly referred to other possible causes of the condition, for example, prior to the diagnosis of thalassaemia, E had caught a chill and suffered

convulsions and that E was conceived whilst Mrs. Z. was taking the pill.

In 1984 the family visited Pakistan with E. He was transfused just before leaving and the family returned before the next transfusion was due. In Pakistan visits were made to hakims but a cure was not found. Mr. Z. wanted to send a leaflet on thalassaemia to one of the hakims he had met but was not hopeful as he had listened to other parents at the thalassaemia support group meetings relating their unsuccessful attempts at looking for a cure in Pakistan. The couple also visited shrines to pray. On their return from Pakistan the family moved to a nearby city where Mr. Z. began to work part-time in a local Pakistani shop.

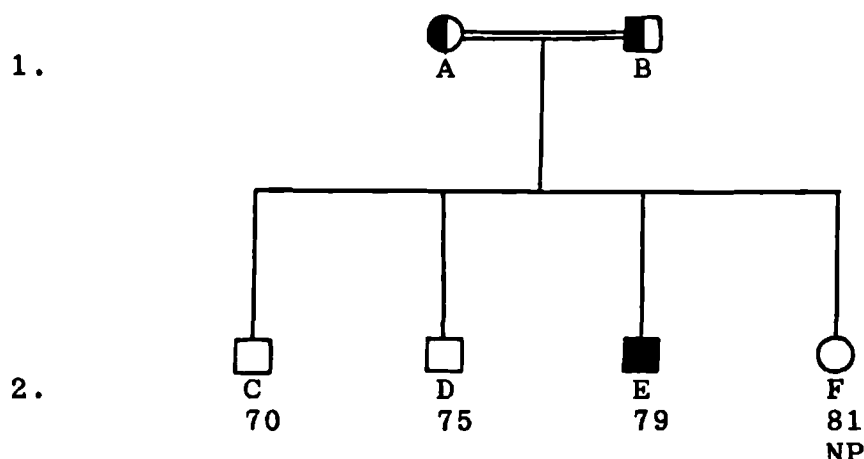
The couple were not planning any more children. In 1984, when the family made another visit to Pakistan, Mrs. Z. had the I.U.D. taken out which had been inserted after E's birth, five years earlier. She did not want any complications to develop with the I.U.D. whilst she was in Pakistan. However she became pregnant during that three month period. Mrs. Z. visited a shrine to pray for a healthy child. The pregnancy remained untested and the baby (F), born in 1985, is healthy. The couple had known about prenatal diagnosis from the support group meetings. On their return from Pakistan, they had felt it was too late to have the test. Also, at that stage the issue of whether to utilise the test had not been resolved:

the couple knew of cases where visits to a shrine and prayer had resulted in healthy children; they were also aware of a case of misdiagnosis and Mrs. Z. was afraid of the actual procedure and after-effects. Her fears were based on experience of having an epidural when E had been born and she said she still felt a cold sensation at the point of entry of the needle. However, she said that she would consider chorionic villus sampling now, as she had recently spoken to a woman who had undergone the test and told her it was simple and painless.

Mr. Z. felt it was better to avoid conception than to deal with an affected child or to have a termination. However, he said, one cannot predict what one would do as one's decision at a given time is dependant on a number of differing factors. Mrs. Z. has had the I.U.D. reinserted and the couple do not plan to have any more children. She did not want to be sterilised as she believed the operation had after effects.

A few months after their return from Pakistan, in early 1985, E had a bone marrow transplant. The operation was successful and E no longer requires transfusions. When I met the couple in 1986, it was the first time I had seen Mr. Z. smile since my first meeting with him in 1981.



Family S

Couple S. are first cousins and originate from villages in different parts of the Punjab. Mr. S. (B), aged 38, born in 1951, came to England 22 years ago in 1967 and has been doing shift work in a factory since. He studied to the eighth grade in Pakistan and speaks a little English. Mrs. S. (A), aged 39, born in 1950, came to England in 1979, nine years after the couple were married in Pakistan. Mrs. S. has not had any formal schooling but was taught to read Arabic and is able to read a little Urdu. She does not speak or understand English. The family live in a small town twenty miles from city Z where E is the only thalassaemic child.

The couple's first three children (C, D and E) were born in Pakistan in 1970, 1975 and 1979 respectively. The third child has thalassaemia major. He began not to thrive in Pakistan and frequent visits to the doctor did not result in any diagnosis or improvement in health. E was ten months when Mrs. S. came to England in 1979. The couple had wanted to be

reunited earlier but Mrs. S. had had problems obtaining entry. A number of different G.P.s were seen before E was diagnosed as thalassaemic at eleven months. The couple were also told that the condition was caused by their being cousins. They remember information about thalassaemia being given to them in English but say that they did not understand all of it. They did understand that the condition could recur in future children but do not remember any mention of prenatal diagnosis.

Mrs. S. had not used any contraception since her marriage. In Pakistan she had not felt it necessary for her to consider it. In England, at the time she did not know anyone well enough who would have prompted her to think of it, nor did she know what was available and where it was available. Contraception had not been discussed at the hospital when E had been diagnosed. Mrs. S. became pregnant in March 1980. She was worried about the pregnancy but was not aware that anything could be done. When she was three to four months pregnant a social worker visited and told the couple that prenatal diagnosis was available. She also told the couple that the test would cost them £600 and that Mrs. S. would need to stay in London for one week. The couple wanted the pregnancy to be tested but as they did not have enough money, the pregnancy continued without being tested. I am not entirely sure of the cause of this information, since prenatal diagnosis is available free of charge on the N.H.S.. However, the figures

mentioned by the family were those for private patients having prenatal diagnosis for thalassaemia at Kings College Hospital, London (the only other centre in the U.K. where it was done for thalassaemia at the time). It seems that the social worker must have phoned this hospital and told them a Pakistani lady wanted prenatal diagnosis, that the respondent at the hospital assumed she was a Pakistani national, and so quoted the private patients' figures. Thus the list of possible types of failure of communication is lengthened.

Fortunately the child, (F), is not affected. After this birth the couple used contraception for three years and do not want any more children. However, presently they are not using any form of contraception. Mrs. S.'s menstrual cycle has become very irregular, and although she is only thirty seven she believes she is now too old to conceive.

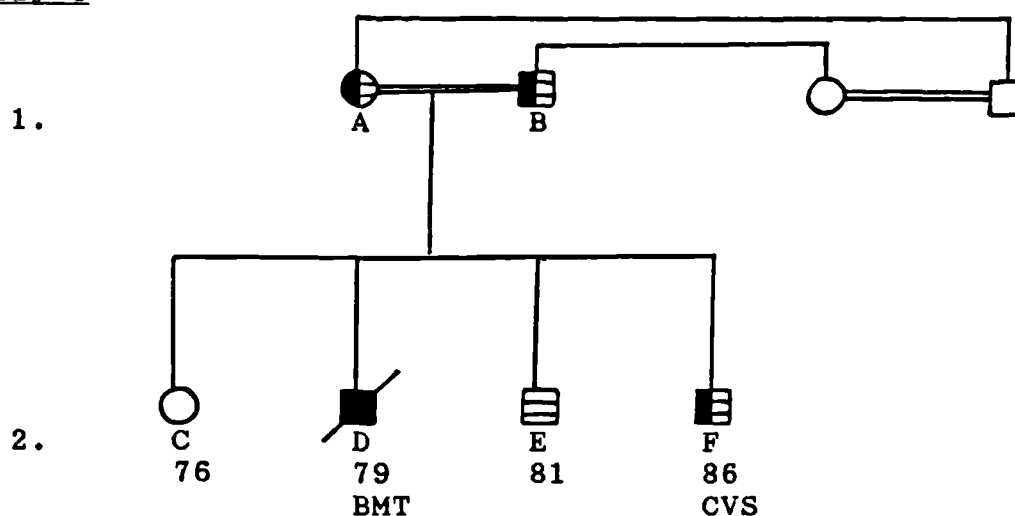
The couple find it taxing to deal with E's treatment. When Mr. S. is working on night shift and supposed to be sleeping during the day he takes E to the hospital. Sometimes he has to take time off work to do so. If he is working during the day Mrs. S. has to take E, leaving the other children in the care of neighbours. She often walks to the hospital as she is not confident travelling by bus. Taxi fares are expensive and not refunded by the hospital.

Mr. Z. said he had learnt a lot from the thalassaemia support

group meetings, to which he was sometimes accompanied by a friend who drove him to city Z. Mrs. Z. had not been able to attend due to the distance and the need to look after the children. However, she said she learned from and felt comforted by the home visits. Both said they were not able to talk freely with the doctor. They felt the doctor continues to speak without checking to see if what he is saying has been understood.

Mrs. S. and other people she knew, had watched a TV programme for viewers of Asian origin, broadcast in Urdu, on which it had been said that thalassaemic children live only till the age of twenty. She says she had spent the whole day crying. It was not made clear that the statement was only true in particular circumstances.

The couple have not visited any hakims or pirs for advice. They do talk openly about the condition with friends and family members. The couple feel it is best to know the carrier status of their children as soon as possible and that it is better to prevent conception than to terminate a pregnancy but, having conceived, it is better to have prenatal diagnosis and to abort the affected fetus than to have a thalassaemic child.

Family U

Mrs. U. (A), born in 1956, is 33 years old. She has lived in England since the age of five with her parents and siblings. The family are originally of a rural background. She has been through the English school system up to the age of sixteen and is fluent in both English and Punjabi. In 1974, at the age of eighteen, she married her first cousin once removed (B) who had come from Pakistan for the marriage. Mr. U. is literate in Urdu and speaks a little English and since coming to Britain has worked in a factory. At the beginning of the research the couple lived in a back-to-back terraced house in the inner city. Mr. U.'s sister is married to his wife's brother.

The couple's first child (C), born in 1976, is healthy. The second child (D), born in 1979, had thalassaemia major and the third child (E), born in 1981, suffers from cystic fibrosis. E requires four types of medication per day plus daily

physiotherapy and intermittent hospital treatment. All the children were delivered by caesarian section.

The thalassaemic son was well until he began to take solids but then became weak and pale. Mrs. U. visited her G.P. and the Children's Clinic twice but to no avail. She was told there was nothing to worry about, but she says she knew there was something wrong with her child. It was only when staff at the clinic arranged an appointment at the Children's Hospital that thalassaemia major was diagnosed. Mrs. U. has felt extremely angry with her G.P. and the clinic for treating her like a neurotic mother. "I feel like sueing them," she said.

The first meeting with the couple was in the hospital. The only knowledge they had about the condition was that it was a disease of the blood and that regular blood transfusions and injections of Desferal were required for a lifetime. They were not aware of prenatal diagnosis. At this initial meeting Mrs. U. appeared very confident and seemed to be coping exceptionally well with two chronically sick children. But towards the end of the conversation she began to cry saying the strain was too much for her at times.

The subsequent meetings took place in the home. On learning the diagnosis, Mrs. U's mother had suggested alternative causes for the condition and that Mrs. U. should visit a holy shrine in Pakistan to pray for her son's health. Mrs. U., who

left Pakistan at the age of five, accepts the medical diagnosis and prognosis for the condition and saw no particular benefit in visiting a shrine for this purpose.

When discussing prenatal diagnosis Mrs. U. was emphatic in her views. She felt that termination was wrong, on religious grounds, that after conception had taken place there should be no interference. The couple had already decided they would not have any more children.

As Mr. U. was at work during the day he was not always present at the meetings, but when I or he wished to discuss something the necessary arrangements were made. Mrs. U. always passed on information and discussed it with her husband. The couple gained an understanding of the genetic implications of the disease and the specific cause. Mrs. U. asked if there were other families with thalassaemic children in the city and expressed a desire to meet them. She attended the thalassaemia society meetings regularly.

When bone marrow transplantation was discussed, the couple decided to go ahead with the procedure despite the risks. They felt that present treatment for thalassaemia was so burdensome that they did not want to deny their son the chance of a complete cure. Their eldest daughter, C, was found to be a compatible donor for D. Soon after this the couple lived separately for personal reasons. It was made apparent

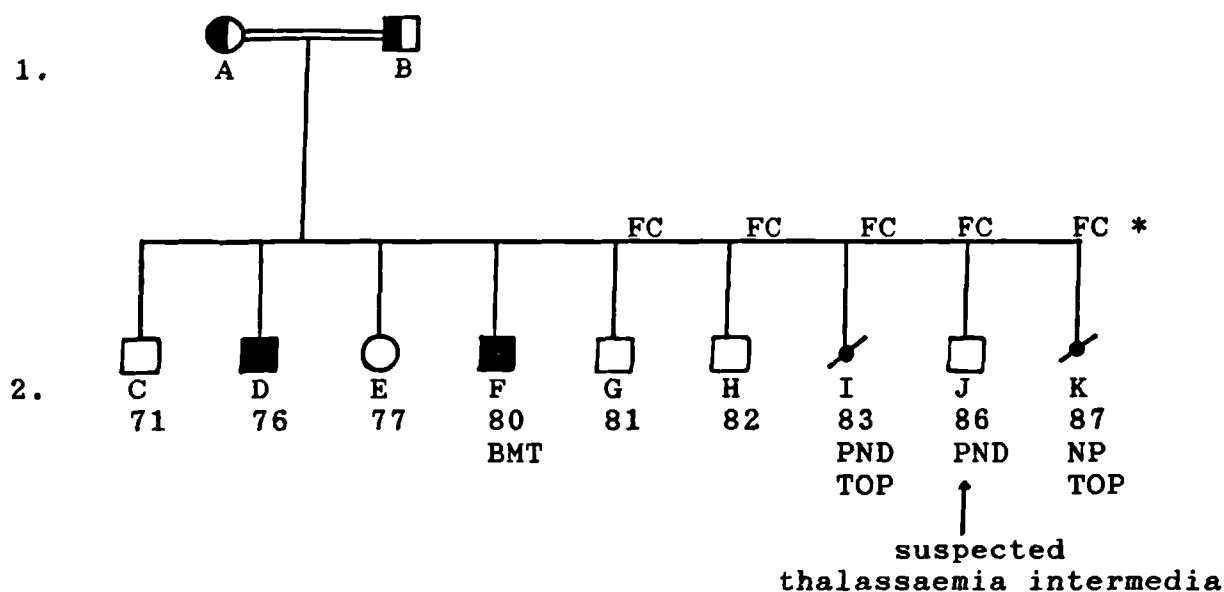
to me that the problems were not due to negative feelings towards the ill children. The eldest daughter, C, who was visiting her paternal grandparents in Pakistan, remained there and Mr. U. followed. Mrs. U. asked if she could donate her own bone marrow. For the tissue typing Mrs. U. and her son, accompanied by me, travelled to London, where we stayed overnight with a member of the Perinatal Centre staff team. The bone marrow was compatible and the operation was carried out, for which Mrs. U. stayed in London with her son at the hospital. For these months Mrs. U. had had to make arrangements for E, who suffers from cystic fibrosis, to be looked after by members of her family. The operation was not successful and D died a few months later. During this period Mrs. U. received a lot of emotional support from her family.

The couple were reunited after the death of their son and made a fresh start in the South of England. Mrs. U. immediately visited the Perinatal Centre at U.C.H. to discuss the risks of thalassaemia major and cystic fibrosis if she became pregnant. On being informed of a 44% risk that any future child would have one or both of these conditions she decided she would not become pregnant until C.V.S. was available for cystic fibrosis as well as for thalassaemia. However in 1986 she became pregnant accidentally. Despite her previous views on termination of pregnancy Mrs. U. had no qualms about deciding to terminate this pregnancy immediately. The suffering her son had endured, especially in the last two



weeks prior to his death, had affected her profoundly. She felt it was better to terminate the pregnancy than to risk having another child with either thalassaemia major or cystic fibrosis.

However, at this stage Mrs. U. received a letter from the Perinatal Centre at U.C.H. informing her that prenatal diagnosis had become possible for cystic fibrosis that week, due to discoveries made at St. Mary's Hospital. She underwent tests for both diseases and delivered a healthy boy (F) who is heterozygous for both conditions. The couple do not plan to have any more children and were ecstatic about their healthy baby.

Family Q

\* Sterilized

Mr. Q. (B) is aged 44 years. He was born in a village in 1945 in Pakistan where he studied to the eighth grade. He came to Britain in 1966 at the age of nineteen and worked in a factory in an industrial city in the north of England. He has a working knowledge of English. He returned to Pakistan in 1969 to marry his first cousin and returned to England in 1970 to live and to work whilst his wife remained in Pakistan, living with Mr. Q.'s family.

Mrs. Q. (A) is 34 years old and was born in 1955 in the same village as her husband. She has not received formal education in a school and hence is not literate in Urdu. However, she was taught to read and recite the Qur'an in Arabic. Presently she does not speak or understand much English. The couple use their mother tongue, Punjabi, in the home.

Mr. Q. made frequent trips to Pakistan whilst his wife was living there and in 1971 a healthy child (C) was born. In 1975 the family were united in England and lived with Mrs. Q.'s aunt. Mrs. Q. was pregnant when she arrived in England and gave birth to a son (D) in 1976. D was unwell from birth and kept in hospital for three months where Mrs. Q. visited every day. On discharge D was still unwell and his condition undiagnosed.

In the meantime Mr. Q. had moved to another city in search of work where his wife and two children joined him to live in rented accommodation. Repeated visits were made to their G.P. on behalf of D and different medication prescribed on each occasion. A lot of resentment is felt by Mrs. Q. towards this G.P. as she felt he was dismissive of her worries. It was by chance when a different G.P., working in the same surgery, suggested a blood test that thalassaemia major was diagnosed. At the time of diagnosis D was 9 months old and Mrs. Q. was five months pregnant with her third child. The couple remember being told that D would require regular blood transfusions and reference was made to what they understood to be certain "germs" as being the cause of the condition. It was only after the birth of the third child (E) that they felt the paediatrician spoke to them in any detail about the condition. On referral to the paediatrician they remember being told, through an interpreter, that the only treatment was regular transfusions for a lifetime, that this condition was caused by

their being consanguineous partners, that it could recur and hence they should not have any more children. They do not remember any specific advice being given on contraception. Since the birth of D the couple have tried the I.U.D., the pill and the condom but have found each to be unsuitable.

The couple have felt desperate regarding their necessity for adequate contraception as the staff at the family planning clinic became increasingly irritated with Mrs. Q.'s need for an alternative contraceptive method. At this time the couple held the belief that, in principle, it was religiously acceptable to use contraception to ensure at least a two year gap between each child to safeguard the mother's health. They did not consider sterilization to be acceptable.

Despite trying not to conceive Mrs. Q. became pregnant for the fourth time and gave birth to a thalassaemic child, F, in 1980. The child was first transfused at six months. A short while after F's birth the whole family returned to Pakistan. The couple hoped that they might find a cure in alternative forms of medicine, although the paediatrician had advised them not to go. Mr. Q. is an only son and he felt duty bound to take the children to visit their elderly grandparents. In Pakistan, despite many visits to alternative practitioners no cure was found and within a few months D died as he had become technically difficult to transfuse. Distressed, the family immediately returned to England and resumed treatment for F.

In 1981 and 1982 Mrs. Q. had two unplanned pregnancies. Both resulted in healthy boys, G and H.

Mrs. Q. became pregnant again for the seventh time in 1983. By this time the couple were aware of prenatal diagnosis through discussions with me, and from the thalassaemia support group meetings. Prenatal diagnosis was requested and the couple were to travel to London and return the same evening. At the station the couple found that the travel warrant issued by the social services, as Mr. Q. was unemployed, was invalid for the early train even though the time they must travel had been clearly stated. They waited at the station for two hours for the next train. When they arrived at U.C.H. the medical team was ready to disperse, having finished the other cases and given up hope of the couple arriving. However, the diagnosis was carried out but the limitation on time meant not only that there was not adequate time to counsel the couple and to put them at ease, but that the procedure was done hastily. The stress of the day was further increased by the fact that Mrs. Q. began to bleed (probably due to the haste in carrying out the procedure). It proved necessary for her to remain in the hospital for two nights, but Mr. Q. was told he could not stay in the hospital, so the Perinatal Centre staff tried to find him a cheap hotel, but there were no funds to pay for it. A systematic solution to the problem of overnight stay for the spouses had not been resolved and Mr. Q. was eventually given a camp bed in his wife's room in the hospital. The couple had

to arrange over the phone for their five children to be looked after at home, which also meant that they were not able to keep the incident private. The one-day travel warrant was invalid for the return journey, so a new ticket had to be bought from the personal income of the Perinatal Centre staff. A visit was made to the home between the test and the result, when Mrs. Q. still complained of feeling unwell. The fetus was found to be affected and termination of pregnancy was carried out in the couple's home town.

Bone marrow transplantation had been discussed at a thalassaemia society meeting. The couple requested it for F, and one of his brothers proved to be a compatible donor. The procedure was successfully carried out in London, and F no longer needs transfusions.

The couple's problems with contraception had never been resolved, and in 1986 Mrs. Q. became pregnant for the eighth time, by which time the family had moved to another city. Prenatal diagnosis was again requested, and once again the fetus found to be affected. However the couple decided against termination of pregnancy. Mrs. Q. said she intuitively felt her baby would be healthy. The successful bone marrow transplant probably also influenced the couple's decision, in offering a possible alternative if the child was thalassaemic. The couple are very happy about the birth of this child (J) but panic when he has even a minor illness for

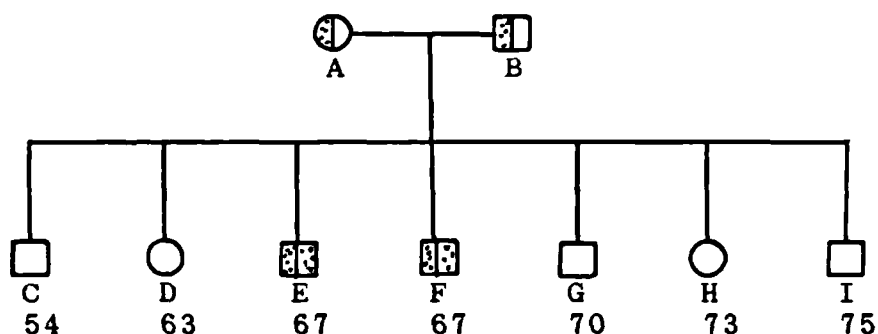
fear that he may be thalassaemic. The child has not yet required any transfusions. Medically it is suspected that he may have thalassaemia intermedia.

In 1987 Mrs. Q. became pregnant for the ninth time. She had this pregnancy terminated in the early stages without prenatal diagnosis, and has also been sterilized. Mrs. Q. explained that Mr. Q. had said he would abide by which ever decision she made and she felt she had no choice in the matter. The decision to terminate was due to a culmination of pressures. Particularly with their reproductive risk the couple had not planned to have so many children. Mr. Q. had been made redundant a few years ago and despite having moved to another city in search of work had had no luck. Mrs. Q. did not want to undergo prenatal diagnosis again as she had found the process very stressful and the procedure degrading as, at the time, chorionic villus sampling was performed transcervically and by a male obstetrician in London. She was not aware that the transabdominal procedure was now being offered nearer her home.

Both Mr. and Mrs. Q. are a religious couple and members of a philosophical order within Islam. In their home, although small, they keep one room specifically for prayers, meditation and religious gatherings and meet regularly with other like-minded people in the city. The couple have a family pir in their city from whom they seek counsel.

I first visited the family in their home in October 1982. After discussion about the condition they said it was the first time they had actually understood the cause of the disease. Up to this time they had felt very confused, and that was why they had sought alternative treatment in Pakistan. They had known that there was a risk in each pregnancy of having a thalassaemic child, so Mrs. Q. had informed her G.P. of every pregnancy and attended the antenatal clinic, but no mention had been made of prenatal diagnosis until the thalassaemia society meetings in 1982. When deliberating whether prenatal diagnosis and termination of an affected fetus was morally acceptable they consulted their pir who advised them that with a condition that causes so much hardship it would be acceptable. The couple felt that they had had to decide on a number of occasions which was the lesser of two evils. They said prenatal diagnosis ought to be available but there should be no compulsion in using the service. They felt they had made their decisions according to their own situation and belief and other people ought to do the same.



Partially studied familiesFamily J

Couple J. came to England 32 years ago in 1957. Mr J. (B) arrived ten months before his wife (A) and one son (C). Mr. J. is 61 years old and his wife a few years younger, is in her late fifties. They live in a rundown part of the inner city in a small terraced house. The conditions in the home are very overcrowded as they now have seven children, the youngest being fourteen years old. Couple J. are of rural origin with little formal education. Mr. J. speaks a little English and has worked in a textile factory and as a taxi driver. He was seriously ill in 1981 from which he has not fully recovered. He has also not been able to find suitable work since then.

Mrs. J. speaks only Punjabi, is literate in Punjabi and is able to read the Qur'an in Arabic. Living in an area populated by people originally from Mirpur District, she performs the voluntary and respected task of teaching the neighbourhood children to read and recite the Qur'an. The younger women living nearby also come to her for advice on

religious matters. Mrs. J. spends much of her time at home due to ill health. She uses the audio-cassette to listen to recordings from her pir on religious topics to increase her knowledge.

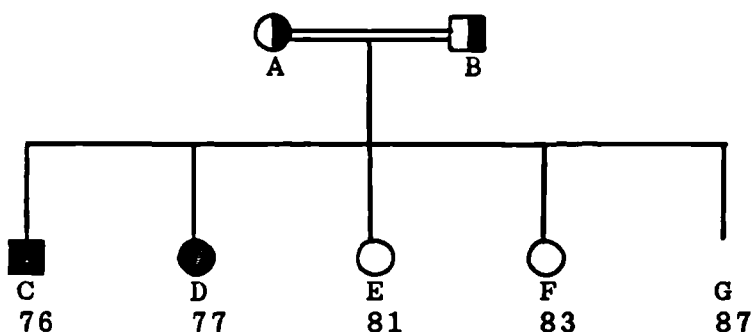
The third and fourth children (E and F) suffer from thalassaemia intermedia. There is an age difference of eleven months between the two children. The younger of the two, F, was diagnosed before E at the age of two years and one month, when he presented at the hospital with severe anaemia and a large spleen, which was subsequently removed. F sometimes requires a transfusion but not on a regular basis which the family find confusing as E, who has the same disease has regular transfusions. F's growth is stunted and he has the thalassaemic facial changes. His eyes are always yellowish in colour which the couple think may be caused by the fact that he suffered from dysentery as a child. The condition has caused him a great deal of distress psychologically. He is socially withdrawn and used to dread going to school where he had been placed at the lower end of the E.S.N. (educationally sub-normal) scale. His lack of achievement at school was also attributed, by the school staff, to partial deafness, but the possible link between deafness and apparent lack of intelligence has not been resolved. This information on F was obtained from his parents as F would not enter into a conversation with me himself. Within the home F is spoken of with affection and was said to be talkative but only with his

family. On the other hand, his brother, E, suffers very mildly with thalassaemia. E presented and was diagnosed as having thalassaemia intermedia at ten years and two months after a road accident. Prior to that he been healthy and even now looks like any other young man in his early twenties. He does, however, require blood transfusions and uses the pump to administer Desferal himself. He was happy at school and is generally quite confident. Both E and F attended the same school where E took on a protective role due to the difficulties his brother was facing.

The first meeting with the family had been at a thalassaemia support group meeting where Mr. J. had attended with E and F. They had responded to a letter as the family did not have a telephone at the time. Mrs. J. has never attended any of the meetings due to poor health and the need to be at home when the other children come home from school. Subsequent meetings took place in the family's home. Discussion, however, has been hampered by the lack of space and the number of people coming in and out of the room as the front door opens into the sole sitting room. Only on one occasion was I able to have a lengthy conversation with Mrs. J. alone. This time was spent discussing the inherited nature of the disease, the availability of prenatal diagnosis and the worries and sadness Mrs. J. felt about her two sons, particularly F. Mrs. J. has a mistrust of doctors who, she feels, repeatedly ask for blood samples despite her ill health but offer little help when she

feels worse after the samples have been taken.

Recently, the eldest son (C) has married his cousin from Pakistan and is presently living with his parents. At the last meeting with the family, couple J. and a number of children were in the room. Whilst informing me about E and F's health Mr. J. wanted me to explain, once again, the cause of thalassaemia, even though he had attended all the support group meetings. He said, "To be honest I have still not fully understood". This proved to be a learning experience for me in terms of the constraints and dynamics involved in the imparting of information on inherited conditions (particularly without teaching aids). Sometimes, it is only in the comfort and safety of their own homes that parents, who find it difficult to grasp the nature of inherited disease, will feel comfortable enough to ask for repeated explanations.

Family V

Mr. V. (B), born in 1950, aged 39 years, came to Britain in 1967 at the age of seventeen from Mirpur District. He came to join his father and brothers who were living and working here. Since his arrival he has worked in textile factories in a small town ten miles from city Z. In Pakistan he had studied to the seventh grade and is literate in Urdu but speaks very little English. The year before coming to England he had married his first cousin and made repeated visits back to Pakistan. In 1977 Mrs. V. (A) joined him in England with their first child (C), who suffers from thalassaemia major. Mrs. V. studied to the sixth grade in Pakistan, is literate in Urdu and speaks no English.

C, who was born in Pakistan in 1976 was unwell from birth and was diagnosed as thalassaemic at six weeks when Mrs. V. had taken the child to a paediatrician in Islamabad, the nearest city to her village. Mr. V. went to Pakistan when he learnt their child was unwell and to speed up the process of getting his wife to join him in England. The couple had found the transfusions to be very expensive and the distance to

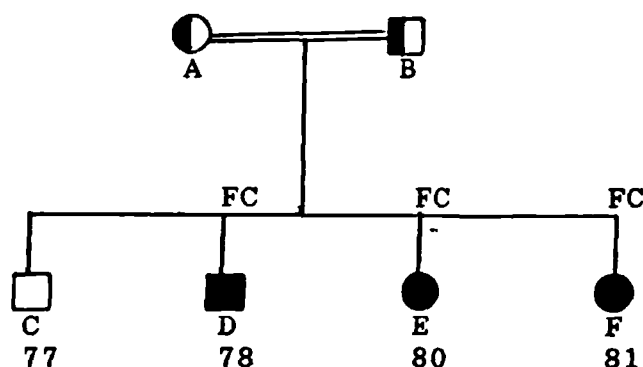
Islamabad very far. They had not understood in Pakistan that the condition was inherited. On arrival in England, C began to receive regular transfusions and Mrs. V. was already pregnant with her second child (D). The doctor discussed family planning with them with the aid of an interpreter. They do not remember being told about the inheritance pattern of thalassaemia. However, they do remember being told that all their future offspring would suffer from the same condition. They were also told that the cause of the condition was because they were cousins and that if they had not been cousins this would not have occurred. D was born in 1977 and also has thalassaemia major.

Four years later, in 1981, Mrs. V. gave birth to a healthy daughter. She had not informed her G.P. or the antenatal clinic of the pregnancy. In 1983 she gave birth to another healthy daughter, again she had not informed anyone of the pregnancy. The couple explained that with two unhealthy children they had wanted a healthy child. Many of the couples that they knew were cousins and had healthy children, so they had disregarded the doctor's words and had decided to have further children. They added that it is frightening to disregard medical advice but the doctor's explanations did not make sense. They felt that the fact that their subsequent children were healthy confirmed that the doctor was wrong. The two healthy children are daughters and the couple would also like a healthy son.

In 1986, when I visited the family at their home Mrs. V. was pregnant again. The couple did not know that prenatal diagnosis was available. It was not possible at this meeting to discuss their feelings about prenatal diagnosis, in any detail, as some of their relatives were present and the small children required attention. Their doctor at the hospital was informed about the availability of the test and that couple V. may wish to use it.

The couple said they worried about their eldest two children's future and wanted to know more about bone marrow transplantation. They, along with their relatives, also wanted to discuss the significance of consanguineous marriage in causing disease, as there seemed to be so much debate about it and they had relatives who were contemplating marriage with their cousins.

Mr. V. had only been able to attend one support group meeting as he did shift work. When he did attend he was accompanied by a family friend. Mrs. V. could not attend alone because of the distance (10 miles) and because the other children needed to be looked after.

Family C

Mr. and Mrs. C. are first cousins from different villages in the Mirpur District. Mr. C. (B), born in 1954, aged 35 years came to Britain to join his father at the age of thirteen. He went to school until he was fifteen and worked in a factory until 1986 when he became unemployed. His employment was affected by the fact that he took time off to see to the needs of his three thalassaemic children. He has been unable to find work since then. He speaks English fluently but feels more comfortable conversing in his mother tongue, Punjabi. Mrs. C. (A), aged 29 years and born in 1960, came to Britain immediately after her marriage in Pakistan in 1976. The family live in a city twelve miles from city Z. Mrs. C. gave birth to her first child (C), who is healthy, in 1977. Her second child (D), born a year after in 1978, has thalassaemia major. The child was well until 18 months and then began not to thrive. He was taken to the G.P. who immediately sent him to hospital where he was kept for one month and then diagnosed as having thalassaemia major. At the time the couple's third child (E), born in 1980, was five months old. Mrs. C. was



also three months pregnant with her fourth child (F). E and F also have thalassaemia major. The couple said the doctor suggested Mrs. C. terminate her last pregnancy but the couple did not want to because of their religious beliefs. The doctor, however, said that he had suggested prenatal diagnosis with the option of terminating an affected pregnancy. The couple resolved not to have any more children. Mrs. C. felt under pressure from the doctors to be sterilized after her last delivery but refused, as she feared that she may be unwell after the operation and unable to look after her children. She has been using an I.U.D. since and there have been no more pregnancies. After her first child, before the birth of any of the thalassaemic children, Mrs. C. was taking the pill but felt unwell and discontinued this form of contraception. She had not wanted to have children so soon and without time-gaps in between, but she says she was newly-arrived from Pakistan and it had taken time to settle and learn of the availability of family planning services.

The couple had been informed of the inherited nature of the disease and its treatment, but in English, so only Mr. C. had understood. Although Mrs. C. is now able to converse a little in English, at the time of the diagnosis she did not understand English at all. She said initially her husband did not tell her the transfusions would be for a lifetime as he felt that she would be upset, so he carried the burden of the knowledge alone. The couple felt the thalassaemia support

group meetings had helped enormously in learning more about the condition and in meeting people. These meetings were held locally and were the initiative of a health visitor who had asked me to run the discussions whilst she made the practical arrangements to bring the families together.

Prior to the meetings the couple had known of two families in the same situation, but as they said, "not from their part of the world" (meaning no families from their part of Mirpur District with whom they felt a lot of affinity). Mrs. C. says she had wanted to know more about the condition but prior to the meetings did not know who to ask. The couple also said that despite being told, the information took a long time to sink in.

Mrs. C. commented that the language factor and cultural background of the person giving information and support has been very important. She stated that "We have a very kind health visitor who wants to help, but due to the language barrier it's difficult to communicate everything I want to say effectively. Discussing things with people who have the same ways and language and participating in the discussion is just something else altogether".

Families with thalassaemic children in another county in England.

As a result of a talk given to doctors at a regional conference, I was asked by a paediatrician to present the findings of the research to a group of her colleagues in a city, 40 miles from city Z., in a different county. This presentation was to be followed by a discussion with all the families with thalassaemic children in the city - seven families with a total of thirteen thalassaemic children. One of the families was family Q. who had moved from city Z. This was the first time the families had been brought together, and also the first time they would be discussing their childrens' disease with someone of the same cultural background and in a language they fully understood. On the morning of the meeting I was delayed by three hours due to involvement in a road accident. On arrival, every family but Mr. Q. (who returned later) had remained behind to wait - an indication of their need for the meeting.

The discussion with the families lasted about three hours. None of the families had understood the inherited nature of the disease, its prognosis or the options available to them, including prenatal diagnosis. The issues arising were similar to those in other cities: the isolation, the need for more information in their own language, fears about their child's present and future health, lack of knowledge about the disease and the options available. One mother was terminating every pregnancy through fear of having another thalassaemic child,

another was refusing transfusions for her second thalassaemic child as she felt that transfusing her first thalassaemic child had created the child's need to be transfused for a lifetime. A teenage girl with thalassaemia had stunted growth and skeletal deformity was shy and withdrawn. On being told that there were other people of her age with thalassaemia, she and another teenager said they would like to meet up. As a result of the feedback from this meeting the paediatricians decided to set up a support group for the families.

#### Conclusion: Family studies

All the case studies presented in this chapter illustrate distressing situations. There is no reason to doubt that similar situations exist in other parts of the country. It is abundantly clear that the health services available to the rest of the population are not reaching this group.

A number of common themes emerge from the foregoing case histories: some highlight deficiencies in service delivery and others illustrate the dynamics involved in decision-making particularly relevant to muslim people of Pakistani origin. A knowledge and understanding of both is required by service providers who determine the nature of a service, and by other health personnel who have face to face contact with the families.

## THEMES AND DISCUSSION ARISING FROM THE FAMILY STUDIES

### Community knowledge of thalassaemia

There is no community knowledge of thalassaemia among the British-Pakistani population nor among Pakistanis in Pakistan. Regardless of educational background, the only people aware of the condition or of inherited disease were the medical profession or informed families of the sufferers.

### Understanding of thalassaemia

At the beginning of the study only two families understood the inherited nature of the condition and its implications for further reproduction. One was couple O., where Mrs. O. had a thalassaemic sibling, but only learnt of the inheritance pattern after she herself had given birth to a thalassaemic child. Couple P. learned of the nature of the condition from doctors in Pakistan. The rest of the parents either did not remember information given to them, or had not understood what they had been told, were sometimes misinformed, and in most cases were indirectly blamed for causing the disease by having a consanguineous marriage.

### Isolation of the families

The families felt very isolated: only a few couples in city Z. were aware that there were other thalassaemic children in the city. None had been introduced to one another and no initiative had been taken to bring the families together.

There appeared to be a general opinion among health professionals that British Pakistani women would not be allowed, by their husbands, to attend the meetings, and that there was stigma attached to handicap which would limit the success of any such initiative. Most of the couples thought theirs was the only child with the condition. The thalassaemia support group meetings helped greatly in combating isolation and in the development of informal supportive networks, as well as being a focal point for learning, discussion and exchange of information and experiences. Once health professionals in other cities learned about the success of the group, an effort was made by some to start their own local group.

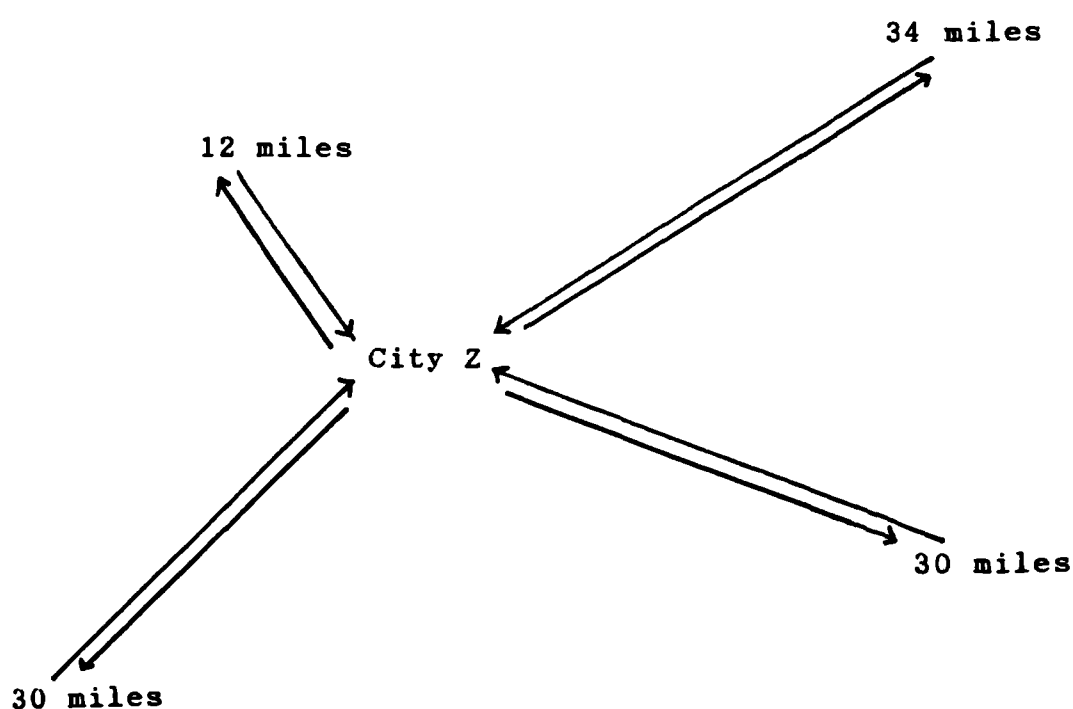


Fig. 3.1: Distances travelled by families to attend thalassaemia support group meetings in city Z.

The lack of stigma

Parents have tended to be very open about their child's disease. None of the parents in the intensively and partially studied families blamed themselves for their child's disease nor were any mothers blamed for bringing the disease into the family. The child's disease was accepted as a part of their fate and the tendency was to look forward. "Why me?", was a question I did not hear expressed.

The child's condition was discussed openly with other family members both in England and in Pakistan. There has been a willingness to meet other people in the same situation and to exchange information. This lack of stigma and lack of blame directed toward parents themselves may be due to Islamic beliefs which parents expressed: that difficulties are an inevitable part of life and some are willed by God to develop an individual's inner strength and capacity to cope with life; that God never puts a greater burden on a person than he or she has the capacity to bear. Some parents said that they sought solace from the fact that they had done as much as they could, given their circumstances and degree of knowledge.

The lack of blame directed specifically at mothers (see pp.278-284 for a fuller discussion) may be due to the fact that many of the marriages were consanguineous and therefore, to blame one's wife's family for "bad blood" would be akin to blaming one's own family.

The lack of stigma among families has resulted in their active participation in support groups which would not have been possible if stigma had existed.

#### Fatalism

During the course of the research the opinion that Muslims were fatalistic and left everything to God (implying they did not take responsibility for decisions or use their initiative) was often voiced by health professionals. This assumption was also used by health professionals to justify not starting self help groups for this population. As mentioned earlier, the meetings were well attended by both parents. Other examples of using initiative were shown by couple R. and couple P. in the efforts made to have blood samples tested from family members in England, the U.S.A., in cities or villages in Pakistan in the hope of finding a compatible donor for bone marrow transplantation. The two couples differ considerably in terms of social background and education but the unifying factor is that both sets of parents had understood the information provided on bone marrow transplantation and desired the operation for their child. The efforts that parents have made to ensure having a healthy child, for example, visiting pirs, and seeking alternative treatment are a further indication of parents using thier initiative to improve their child's health rather than playing a passive role.



One's fate (taqdheer) in the muslim context is seen to be those things over which one has no control such as one's birth, the family one is born into, in fact any event that one cannot predict and control. A parallel concept to taqdheer is "tadhbeer" which is that which a person wills to mould his given fate. So, a parent saying "It's all in God's hands" does not necessarily mean I have no control. For a parent who is seeking the best for the child, it means that with the information available to me I will do as much as I can, within my moral parameters, but then whatever happens I will accept it as being God's will. In psychological terminology this can be seen as a defence mechanism in coping with the decisions that parents have to make. However, it could be that some parents may use these principles to avoid making decisions about treatment and prevention. It is important that the genetic counsellor is able to perceive the emotional state of the parents and discern whether the parent is stating his or her personal view or is fearful of desired change.

All the parents sought further information and were making decisions which were rational to themselves in the light of the information available to them. This is consistent with another well known Islamic principle: that it is incumbent on a person to seek knowledge and not to live in ignorance. Some parents emphasised this principle; it can be a point of discussion between a counsellor and parents to aid parents in dealing with new information and options in a familiar

context.

#### Religious beliefs and their application

Contrary to media projections and popular Western perception of Muslims (see p.282-283 for further details) couples have applied their religious beliefs to themselves as individuals, and do not function within a dogmatic framework. Couples had different views on termination of pregnancy and made different decisions. Many have stated that given the chronic nature of the disease each person must decide for themselves their course of action, depending on their circumstance and their own understanding of their faith. Couples who have visited pirs regard them as highly respected advisers and spiritual guides, but the only authority they have is that vested in them by those who seek their counsel. None of the couples thought of going to the imam at the local mosque for advice.

#### Misinformation

The key tool in genetic counselling is information. It is on the basis of the information given about the disease, the treatment, the risk of an affected child and the preventative options available, that couples are able to make informed choices about their reproduction. By the same token, the main hindrance to genetic counselling is misinformation. The family studies illustrate many examples of misinformation which have affected parental attitudes, behaviour and decision-making. Of the couples who were in a consanguineous

marriage, virtually all were initially told by a health worker that the condition was "caused" by the couple being cousins. This was not only false information, but in the process, a correct explanation of their child's disease was denied to the parents. Citing consanguineous marriage as the "cause" of thalassaemia placed the burden of responsibility on the parents' shoulders and simultaneously away from the health professionals. It also implies that the marriage pattern of the community has caused them to have an affected child and the sole option available to them and the other people in their society is to change that marriage pattern. Coupled with absence of information on the real options available, the result of such communication can only be to produce guilt and despair in the parents.

As consanguineous marriage is highly favoured among Pakistanis, such statements are also seen as an attack on what is viewed as an established and positive social practice. That the statements regarding consanguinity are false become apparent to the parents very quickly, as they are surrounded by couples who are cousins but who do not have thalassaemic children. This in turn produces mistrust of the health professional, whose advice in the future will not be considered sound. These comments have caused strong reactions in the parents. Mrs. O., on being told what she knew to be false information said, "I nearly hit him (the doctor) with my handbag. I didn't, instead I vowed never go back to him".

Mrs. A. on being told that the disease was caused by her consanguineous marriage and that all her future children would be thalassaemic gathered the courage to become pregnant again as she had two thalassaemic children and no healthy children. She knew the first statement to be false and therefore doubted the validity of the second. However, she did not inform her GP or the antenatal clinic of the pregnancy until the final stages, thereby foregoing ante-natal care.

Genetic terminology, such as autosomal recessive inheritance, genes, D.N.A. is complex for anyone other than those who have studied it. To explain the terms is also difficult for someone who has not been trained to explain them in a simple manner to the lay person. The terms are even more difficult to comprehend by someone who has never heard them before. For most of the parents in the study this situation was compounded by the language barrier. In two cases where the medical practitioner has tried to explain the term "genes" it had been understood to mean "germs". If the condition is caused by "germs", the parents' view was that surely with the present state of medicine there must be a cure somewhere. This explains, for example, couple Q taking their children to Pakistan to look for a cure. Subsequently, when the parents realise that the condition is due to inheritance and has nothing to do with germs it confirms for them that they cannot rely on the medical practitioner for information. There is recognition of difficulties in communicating with health

personel due to the language barrier. If interpreters are not available or not used it is natural that health personell are not perceived as a useful source of information, particularly if mistrust also exists. Hence alternatives are sought.

The setting in which information is imparted is also important. The G.P.'s surgery or hospital office is impersonal. It may be the personal space of the doctor but not that of the parents. The fact that the doctor is usually male also adds some distance in the communication as it is particularly the mother who needs to understand the information. When the mother speaks even less English than the father there can be a tendency to address the father only. He may nod as the information is given, meaning that he is listening, but this does not necessarily mean that he has understood. The research showed greatly increased understanding and communication when the mothers and couples were seen in the home by a female of the same linguistic, cultural and religious background. The home is the personal territory of the parents and hence where they feel comfortable voicing their concerns, doubts and anger.

#### Delay in Diagnosis

The delay in making a diagnosis which ranged from one month to twelve months has meant prolonged periods of stress for parents, repeatedly going back and forth to the G.P. or hospital, knowing something was wrong with their child without

any resolution. The distress was further increased by the dismissive attitudes of the doctors to the parents' worries. Mrs. Q.'s child was unwell from birth and kept in hospital for three months, where she visited him every day. The child was discharged without a diagnosis and eventually diagnosed at 9 months. Mrs. J. related how she spent endless nights with her daughter crying:

"I felt so much pain but there was nothing I could do."

Mrs. U. said she felt she was treated like a neurotic mother by her GP and the antenatal clinic staff:

"I feel like suing them."

This is a common problem with inherited diseases, as they are rare and most G.P.s will maybe see only one in a lifetime. Even the most informed G.P.s may be slow in making a diagnosis. However, the present situation leaves a lot of room for improvement.

#### The Role of the G.P.

The G.P. has a pivotal role to play in the existing infrastructure of the medical services. He or she is the first point of contact when there is a health-related problem and also the source of access to more specialist services. The family studies illustrate many cases of how lack of adequate knowledge or training for G.P.s in the field of genetics has led to the imparting of false information with

serious consequences. For example, Mrs. I., who wanted chorionic villus sampling, was told by her G.P. that no prenatal testing existed, hence the pregnancy was continued and she delivered a thalassaemic baby. Another area of concern was many G.P.'s inability to have thalassaemia diagnosed promptly and mothers expressed anger at being treated as neurotic women when they repeatedly returned to their G.P. with an unwell child.

A bad rapport at this initial stage creates alienation between the G.P. and the couple, at a point when the couple are perhaps at their most vulnerable, and when they will be needing to rely on doctors in the future. It is at this crucial stage that a speedy diagnosis should be made, the correct information given sympathetically and referral made to specialist and ancillary support services.

### Contraception

The case studies provide many examples of failure of contraception resulting in unwanted pregnancies. All the mothers in this study were born in Pakistan and the majority spent much of their adolescent years there. Hence on arrival they are not aware of the family planning and ante-natal services in Britain, and newly married women who arrive in England may not initially be close to anyone who could inform them. However, it is generally expected that there will be a child soon after marriage, though there may subsequently be a

desire to have gaps between the children.

Family planning was discussed only with a few couples at the time of diagnosis. This is the most appropriate point at which to ensure that couples receive appropriate contraception, particularly as a "no pregnancy" stage would allow parents to deal with the new-found knowledge on the disease and the options.

Many couples using contraception were finding it to be ineffective, particularly the condom. Frustration was felt whenever a pregnancy had resulted from contraceptive failure. Amongst four couples there have been eleven unwanted pregnancies caused by failure of a contraceptive method. Fears exist among many of the women in the study about the implications of certain forms of contraception and particularly of sterilisation, as they felt it would jeopardize their health. However, four of the eleven women in the intensively studied families did eventually choose to be sterilised, despite the fears. Three of these four women had had a total of ten unwanted pregnancies. The decision to undergo the operation is an indicator of the need not to conceive again given the risks involved and the lack of faith in other forms of contraception.

The case studies show that at-risk families are not receiving appropriate family planning services and information so that



they can use contraception as a way of limiting their pregnancies. The desire to do so is certainly there.

### Prenatal Diagnosis

The family studies clearly belie the notion that "There is no point in offering prenatal diagnosis to muslim families. They will not use it as it is against their religious beliefs." This research shows that in reality this notion, which is no more than an assumption, is used to deny a population access to a service. Utilising a premise so rigid in its affirmation also precludes any intention of providing that service. The family studies show that most families were not aware of the availability of prenatal diagnosis and thus were unable to make a decision about using it.

When making an informed choice, some couples rejected prenatal diagnosis totally, either for religious reasons, or as a result of negative experiences. Some rejected prenatal diagnosis at sixteen weeks but found it acceptable at nine weeks. Others have accepted it both at sixteen weeks and at nine weeks, particularly those couples who have experienced looking after a thalassaemic child over a long period in Pakistan.

Attitudes, perceptions, needs, behaviour as well as technology are always in a state of flux and broad generalisations applied to individual families are of little value. Women who

did not feel it even necessary to discuss prenatal diagnosis as they were so opposed to termination, in later years changed their minds in the light of painful experiences, such as the death of a thalassaemic child. Others, because of mismanagement or the success of bone marrow transplantation have no longer felt the need to use it. A further category of women, although opposed to termination of pregnancy, found it reassuring to have the test to save the anguish during the pregnancy of not knowing whether the fetus was affected.

The availability of chorionic villus sampling has been an important breakthrough, as termination at an early stage of pregnancy means there is less attachment to the baby. As the pregnancy is not apparent to others the termination can be kept private. For some, termination may be unacceptable at any stage of the pregnancy. However, in these cases future advancements in the field of genetics may be beneficial and couples need to be kept informed of these as they arise.

Another development which affected the uptake rate of prenatal diagnosis in the families studied was the availability of the service locally. For most of the women the journey to London was long and arduous, and involved travelling on the underground, often for the first time. The trip also involved an overnight stay and hotel expenses for the spouse. Arrangements had to be made for the children to stay with someone else, meaning that the matter could not be kept

private. For the unemployed, travel warrants had to be obtained before the trip could be made. Chorionic villus sampling performed transabdominally rather than transcervically, as it had been done earlier, was another development which made prenatal diagnosis more acceptable. One of the reasons, among other for Mrs. Q. deciding not to have the test was that she had found it undignified to have it done transcervically by a male doctor.

Some of the issues discussed above are pertinent only to Muslim families of Pakistani origin in Britain. However, parallels can be drawn with families from an entirely different cultural background which show parents reacting in similar ways and encountering the same experiences with a chronically sick child in the family. Such experiences and reactions are not unique to this minority population; this is illustrated by Burton's study (1975) in northern Ireland of parents and their children with cystic fibrosis, a recessively inherited disease that mainly affects people of N. European origin. Burton (chapter 4) states that the diagnosis is met with shock by parents, like the reaction to a greivous accident:

"I can't recall what we were told on that occasion. An awful lot of what he said I lost. I'd already begun to look out of the window. The feeling I had the roof was coming in around us."

"I didn't even know what it was. I thought it was multiple sclerosis. I couldn't remember what it was she had. I kept crying, but I didn't know. The CF trust secretary had to come up and explain it to me."

She notes that denial was the most widespread of the mechanisms used:

"I thought he might be wrong and that there was nothing wrong with the child at all."

"I felt it wasn't it. That she hasn't got it at all, that she couldn't have it."

"I can't really describe how I felt about it at the time. It's a sort of thing you don't think about.....you push it more to the back of your mind and don't think of it."

On the implications of the emotional turmoil Burton states that:

"Not only do they actively not want to know but also they experience a paralysis of thought processes which renders impossible comprehension of anything but the simplest, clearest information."

It is not surprising, therefore, that given this emotional state and compounded by language difficulties, that parents in the thalassaemia study did not "remember" being told anything about the condition.

In Burton's study mothers who experienced little emotional disturbance were those who had not understood the severity of the condition or its inherited nature.

Misinformation on the part of the doctor exacerbates an already tense situation, as dealing with an inherited condition evokes deep emotions. Burton notes that doctors and healthworkers can become targets for the expression of anger.

In Burton's study parents also felt hostile and angry towards doctors. Like the families in the thalassaemia study, these parents experienced scepticism on the part of the doctors and delays in the diagnosis of cystic fibrosis:

"I knew instinctively from birth that he had it. I knew by his appearance that something was definitely wrong. I jumped to the conclusion that it was the disease. I had a row with the gynaecologist because he wouldn't send him for a test. So I asked the GP to send him up to hospital. He said there was nothing wrong and not to be looking for trouble, it would show itself in time."

"I went to the baby clinic every week. She would gain 1lb one week and lose it the next. They said I was fussing unnecessarily. They said there were skinny and fat babies, and I was fussing too much. I went to a doctor and he gave me some stuff and he said, "You're a young mother, are you sure you won't put it in her ear instead of her mouth? It made me feel a fool."

However, Burton found that when parents realised the rarity of the condition their anger subsided:

"We blamed him at the time but after we read the literature we realised he would only see one of these children in his lifetime."

Burton adds that where the doctor made efforts - as was usually the case - to be attentive, seemed concerned, expressed regret or visited regularly he was forgiven. In the thalassaemia study, if there was a delayed diagnosis and the parental marriage pattern was cited as the cause of the condition, these factors alienated parents from doctors and rapport, which requires trust and support, was not established and further effective communication was hampered.

In Burton's study parents acknowledged their need to maintain a good rapport with the doctor:

"Sometimes it makes me very bitter, but I can't just turn round and blame the doctor. I still feel I need him a lot and I can't say outright."

The need to turn to the doctor for advice and information is felt by all parents regardless of background. It remains the responsibility of the doctor to make a conscious effort to dismantle any barriers which may make him or her less approachable.

Burton found that the parents who coped best were those who were well supported either by their immediate family, health visitor or district nurse. The health visitor invariably offered moral support and took the mother's worries seriously and in some cases was instrumental in pressing for a diagnosis and dealing with medical personnel. In the thalassaemia study there were several cases of health workers initiating a blood test when G.P.s had failed to do so. Health workers, other than doctors, have an important role to play in the provision of a comprehensive service. With the changing nature of Britain's population ongoing training courses for health professionals which specifically explore the implications for health service delivery in a multiracial society are essential.

"I didn't appreciate how serious it was. I knew nothing of it. They just said she'd got CF and that was all - unless he thought I already knew, and whenever I came home I tried to remember to tell my husband."

Similarly, this study where doctors have felt that parents have not co-operated with treatment, it is likely that the parents have not understood the disease or the treatment.

The Cystic Fibrosis Trust, a voluntary organisation with support groups throughout Britain, founded in 1964 in London by interested parents and clinicians has been of great benefit to parents of affected children:

"By putting parents in touch with each other, by providing well written, simple explanations of the child's illness, and by funding research into the condition, they diminish "that isolation which accentuates all suffering" (Saunders, 1969)."

The thalassemia support groups, as indicated previously, proved a valuable source of support with regular attendance by mothers and fathers. Cultural assumptions prevented the formation of a support group prior to its initiation as part of this research. Such groups are held to be the norm for the majority population, as indicated by the number of support groups that exist for other conditions. These organisations are the result of a joint effort by medical personnel, health workers and parents, they are not an achievement of parents alone.

## CONCLUSION

The case studies illustrate a clear example of a minority population in Britain which is not receiving an adequate medical service, whether in the form of a comprehensive genetic counselling service or specifically in being offered prenatal diagnosis plus other related services such as family planning. At this early stage when genetic counselling services are not firmly established in Britain, service delivery to British Pakistanis is pervaded by cultural assumptions which result in that service not being provided, rather than services being moulded to the needs of the population.

The language barrier is often cited as a major problem between parents and medical personnel and remains a scapegoat for the development of responsive services in a multilingual society with cultural variations. The distance that a language barrier places between doctor and parent in developing a supportive relationship without linguistic misunderstandings is self evident. However, in the experiences of the families in this study, superimposed on that is the ethnocentric dynamic that the parents are different from the doctor. When this dynamic is verbalised in comments such as "this condition is caused by you and your husband being cousins", placing a greater burden on the parents without an adequate qualifying explanation, then the distance is further increased. If subsequent meetings with the family are still overshadowed by



feelings in the doctor that he or she is dealing with people who are "different" from him or herself then the gap between the two is increased further still. This dynamic of the parent being "different from the doctor" exists even in cases where the doctor may be of the same cultural background as the parents, as a middle class doctor of Pakistani origin, invariably a first generation emigrant him or herself, does not necessarily identify with fellow countrymen of rural origins. So, although one cannot diminish the importance of an awareness of a patient's cultural background, equally important is the medical principle of "First do no harm, to help where you can and to comfort always." These principles are so often overshadowed by the ethnocentric notion that "their culture creates problems", that the real reasons for lack of communication and non-utilisation of services are obscured. An acknowledgement that doctors are unable to carry out their responsibilities due to the inappropriateness of the present service (lack of trained interpreters, counsellors, teaching aids, support groups and so on) would be far more fruitful in tackling communication and interaction than citing parents of a minority culture as being pathological in their emotional responses, behaviour and decision-making. The family studies have clearly illustrated that the parents behaved in quite logical ways given their points of reference, the lack of information and the inadequacy of the service available to them.

A comparison with Burton's study has illustrated the common experiences that parents share regardless of their cultural, religious and linguistic background and pinpoints common issues that parents of children with inherited diseases face. Psychosocial studies are of tremendous value in acting as a probe to elicit information on family and patient experience and their precise needs, thereby providing a sound base on which to develop appropriate and adequate services.

Finally, the family studies illustrate that people's situation, perceptions, attitudes and religious beliefs are specific to them as individuals and families and change over time. Each family has to be treated individually; this is a basic principle of genetic counselling and applies to everyone regardless of their background. The family studies have also shown that generalisations and stereotypical assumptions have dangerous and far-reaching consequences for people's lives.

PART FOUR: CONSANGUINITY-SOCIAL AND HEALTH IMPLICATIONS

# CONSANGUINEOUS MARRIAGE AMONG BRITISH PAKISTANIS

## Consanguineous marriage

A consanguineous marriage is a marriage between relatives who have a common ancestor (Figs. 4.1 and 4.2).

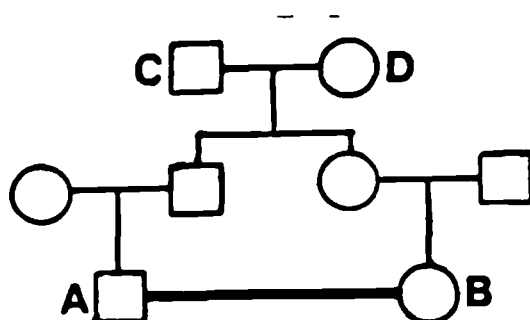


Fig. 4.1: A close consanguineous marriage

A and B are first cousins. They are closely consanguineous as they have a common grandfather (C) and grandmother (D) only two generations back.

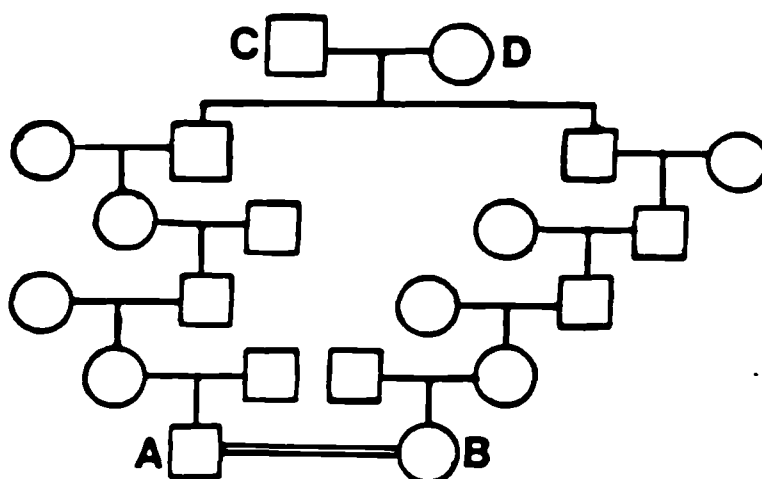


Fig. 4.2 : A distant consanguineous marriage

A and B are consanguineous, but not closely. Consanguinity is only considered close when the common ancestor was only one or two generations back (Fig 4.1). If a common ancestor was many generations back, the consanguinity is considered to be far. Relationships established by marriage (in-laws), where no blood tie prior to the marriage has existed, are not consanguineous.

#### Patterns of Consanguineous Relationships

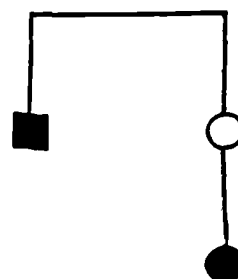
The amount of genetic material that a child inherits from a common ancestor differs according to the degree of consanguinity in the relationship of the child's parents. Some of the patterns of consanguineous relationships are shown in fig. 4.3.

#### Prevalence of consanguineous marriage

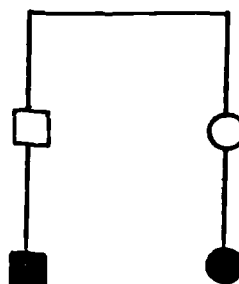
Consanguineous marriage is practiced relatively frequently in many parts of the world (map 4.1).

In Western popular opinion it is generally thought that consanguineous marriage has never been a part of Western culture and the marriage pattern has been associated far more with muslim societies. Consanguineous marriage, though, has been an ancient and well respected tradition, as indicated by the family tree of the patriarchs, Abraham, Isaac and Jacob (fig. 4.4). First cousin marriage is also legal in Britain and many other Western countries. Taking into account the

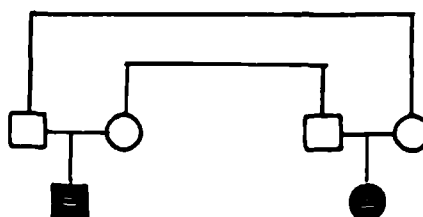
Uncle - neice



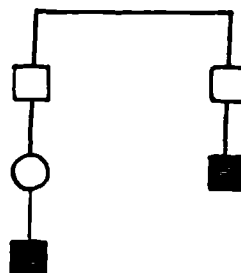
First cousin



Double - first cousin



First cousin once removed



Second cousin

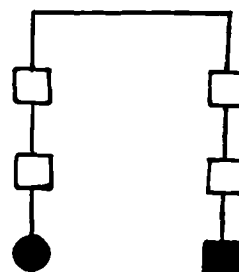


Fig. 4.3: Examples of consanguineous relationships



historical prevalence of consanguineous marriage (Bittles, 1980) a more pertinent question than "Why do people marry their cousins?" would be "Why did people stop the practice?".

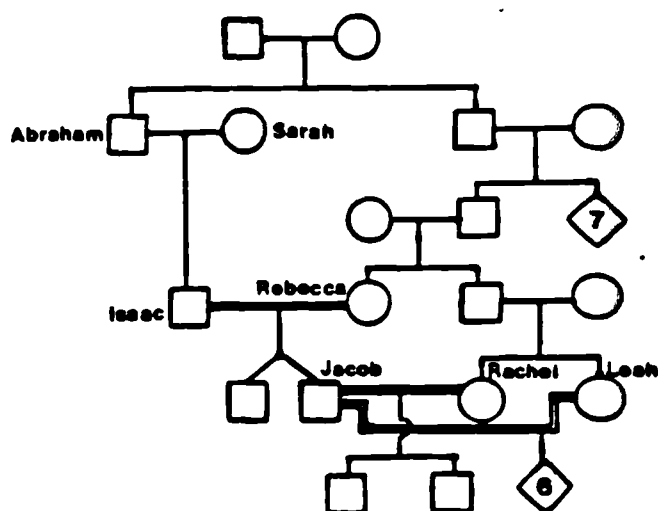


Fig. 4.4: Family tree of patriarchs, Abraham, Isaac and Jacob

In general, however strong the cultural tradition, close consanguineous marriage is more common in rural than in urban areas, and in most countries its frequency is gradually falling as urbanization progresses. Relatively few of the countries most concerned have yet reached the level of development where the need for general genetics services begins to be felt, because as a rule genetic disease begins to be noticed only when the infant mortality has fallen to less than about 60 per thousand.

However, the ethnic minorities in Britain originating from Pakistan is an exception to both these generalizations: on the one hand, social pressures associated with migration have



tended to increase the frequency of consanguineous marriages, while on the other their transfer to a developed society means that their infant mortality has fallen from over 10% to less than 2% in one generation.

In developed countries genetic diseases have emerged as an important cause of childhood mortality and morbidity; particularly conspicuous in groups where the age structure is young and there is a high level of consanguinity, as among the British Pakistanis.

#### The frequency of consanguineous marriage among British Pakistanis

There was a high frequency of first cousin marriage among the parents in this study: in seventeen out of eighteen families the parents were closely consanguineous. Table 4.1 summarises the marriage relationships that were studied more closely.

Group	Number		Relationship				Total known to be married to relatives
	Total	Known to be married	1st cousin	1st cousin once removed	None	Unknown	
Parents of patients	30	30	20*	8	2	0	28/30
Sibs of patients	46	7	4	2	—	1	6/7
Mother's sibs	24	16	8	2	—	6	10/16
Father's sibs	32	27	8	2	8	9	10/27
Total	132	80	40	14	10	16	54/80

\*One double first cousin

Table 4.1<sup>1</sup>: Marriage relationships in 15 families of British Pakistani children with thalassaemia

<sup>1</sup> Source: Darr and Modell, 1988.

Three families were also transmitting a second recessively inherited disorder (two cystic fibrosis and one maple syrup urine disease). The complex pattern of relationships that may occur in one such family are illustrated in fig. 4.5.

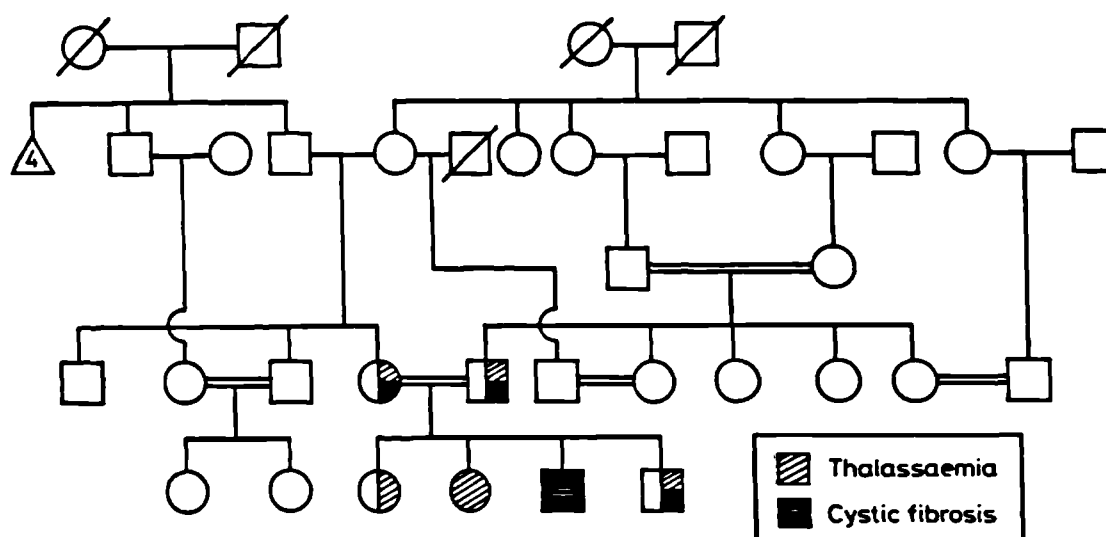


Fig. 4.5: Family tree of a British Pakistani family transmitting both thalassaemia and cystic fibrosis.

Earlier studies have noted a high frequency of consanguineous marriage among the British-Pakistanis (Gatrad et al, 1984). To relate the frequency of consanguinity in the study families to the background frequency of consanguineous marriage among British Pakistanis, I interviewed 101 consecutive women of Pakistani origin admitted to the postnatal wards of the two major hospitals in a city, with the permission of their obstetricians. The study was designed to see if the frequency of consanguineous marriage is changing

with time, so each woman was asked: (1) if she and her husband had been related before marriage, and if so the exact degree of the relationship; (2) the degree of relationship, if any, between her own parents; and (3) the degree of relationship, if any, between the parents of her husband.

When the objective of the study, that is, to gather information on different marriage patterns, was explained, all but one of the mothers agreed to a fuller discussion. The remaining woman gave no reason for declining to be interviewed. Interviews were in the women's mother tongue, Punjabi. In each case a limited family tree was constructed. A full explanation and some delicacy was required, so each interview took an average of twenty minutes. Basic details, such as the mother's age were extracted from the obstetric notes, which also contained a slot for recording consanguinity.

#### Results of the study:

The mothers were mostly young (Fig. 4.6), with an average age of 26 years. All the women were quite clear about their degree of relationship with their husband: 96 were sure of their parents degree of relationship but only 84 were sure of the degree of relationship of their husband's parents. The results are summarised in table 4.2.

The parents:

Fifty five of the women interviewed were married to their first cousins. All four types of cousin marriage occurred: the frequencies are given in table 4.3. No double first cousin marriages were reported in this generation. Nine women were married to first cousins once removed, three to second cousins, and three to more distant relatives (one first cousin twice removed, one second cousin once removed, one third

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<sup>2</sup> Source: Darr and Modell (1988).

<sup>3</sup> Source: Darr and Modell (1988).

cousin). Thirteen were married within the biradheri: in this category some of the husbands were distant relatives and some relatives only by marriage but of the same quom. Only 17 women definitely had completely unrelated husbands but for genetic purposes 30 of the 100 couples could be considered as unrelated.

The hospital notes of above women contained information on consanguinity in only 59 cases and there were 16 errors. Six unrelated couples (three in the biradheri) were described as related, two related couples were described as unrelated, and eight relationships were wrongly described. One mother married to her first cousin confessed to having told the nurse they were unrelated "Because they frown on you and question you if you are married to your cousin".

<i>Woman married to</i>	<i>No</i>
Mother's brother's son	20
Father's brother's son	17
Father's sister's son	11
Mother's sister's son	7
Total	55

Table 4.3<sup>4</sup>: Relative frequency of the different types of first cousin marriage.

The grandparents:

More information was available for the respondent's parents than for her parents-in-law. There is no reason to think that

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<sup>4</sup> Source: Darr and Modell (1988).

there would be differences between the two sets of grandparents, and to eliminate the uncertainties of interpreting the larger number of "unknown and "closely related" responses for the parents-in-law, the more complete and reliable figures for the respondents' own parents were used. In this group of parents there was a maximum of 31 first cousin marriages, with more marriage (24) among the biradheri and more to unrelated partners (30). The four couples whose relationship was unknown are more likely to have been unrelated than closely related, but this has not been assumed here.

That the pattern of inbreeding is not uniform is shown in table 4.4: unrelated couples are more likely to have unrelated parents, while married couples of first cousins more often have closely related parents.

### Conclusions of the consanguinity study

The incidence of first cousin marriage between the parents in this study was found to be 55% but only 30% of the grandmothers were married to their first cousins. This suggests an increasing rate of close consanguineous marriage in this small group, by contrast with some other populations, for example, in Japan, where the frequency of consanguineous marriage is falling as a result of social change and increased mobility (Imaizumi Y, Shinozaki N, 1984). The figures also show an increase above the level of consanguineous marriage in Pakistan. An enquiry among 900 women in hospital in Lahore, Pakistan, in 1983 showed 36% first cousin marriages, 4% first cousin once removed, 8% second cousin and 53% unrelated (of which 25% were in the biradheri (Shami S A, Zahida, 1982). These figures are almost identical with those reported here for the grandparental generation and support the conclusion that the frequency of close consanguineous marriage has increased among British Pakistanis.

The causes of this change were not investigated in this study, but it probably follows from the constraints imposed by migration and the particular kinship networks of this population. It is possible that a similar trend exists among other ethnic minorities in Europe with a tradition of consanguineous marriage, for instance people of North African origin in France and Belgium and people of Turkish origin in Germany and the Scandinavian countries.

The presence and increasing frequency of consanguineous marriage coupled with the young age structure of British Pakistanis has particular implications for the delivery of a genetic counselling service which will be discussed later.

However, in this population, the consanguineous marriage pattern also provides a supportive social structure, especially for women, as well as for men and children, in a patrilineal society. This aspect of the social structure is discussed in the next section as it is necessary to understand it before moving on to discuss the clinical implications of consanguinity. The two facets of consanguinity (social and clinical) combined, provide the necessary background for developing a comprehensive genetic counselling service for all inherited diseases which is appropriate for this population.



THE SOCIAL IMPLICATIONS OF CONSANGUINEOUS MARRIAGE AMONG  
BRITISH PAKISTANIS

A chronic disease like thalassaemia has personal, social and moral as well as practical implications for parents with a sick child, involved both with the treatment and the prevention of the condition. The encounter with thalassaemia is even more awesome for parents who face it for the first time in an unfamiliar cultural environment. They are presented with the diagnosis of a disease which runs in their family that they have never come across in their country of origin. The diagnosis is made by doctors who usually have no understanding of the parents' perceptions of the disease and its causes, or of their response to it. There is usually also a language barrier. The treatment is extremely burdensome, often causing adjustments in the working lives of the fathers, dampening the spirit of siblings and creating a heavy atmosphere in the home. Prevention by aborting an affected fetus is an option which in the first instance is laden with guilt and apprehension as it contradicts the widely held Islamic view on the rights of the unborn child. Resolving this issue forces parents to assess their moral responsibility to the unborn child, their other children, themselves, their spouse and to God. In observing parents with these dilemmas, this research has acted as a "social probe" allowing an insight not only into the practical difficulties that families face, but also into their beliefs, practices and decision-making processes.

The resolution of emotional issues surrounding reproduction, particularly in the context of inherited diseases, usually draws heavily on and tests a couple's bond. The decisions that couples make and the manner in which they make them are indicators of the emotional power relationship between them, and with other members of the family if the issue is discussed openly.

The most striking features of the families in this study grappling with the issue of prenatal diagnosis, was that there was not a single case in which a husband over-ruled his wife's decision on whether to terminate a pregnancy or not; that none of the women were blamed for the disease; that both parents, and in some cases other members of the family, discussed the issue openly with me; that little stigma was attached to having an inherited disease in the family, illustrated by their open discussions in the support group meetings and subsequent visits to each other's homes; that women discussed amongst themselves their experience of prenatal diagnosis and termination at the first support group meeting; and that no interest has been shown in fetal sexing.

All the above points suggest a shared participation and responsibility for the care of the sick child and in deciding about the health of future children. They also illustrate the power of the women over their reproduction which serves as an indicator of their position in the family.

The above characteristics demonstrated by the families in this study were in sharp contrast to B.M.'s experience of Hindu families of Indian origin in London (personal communication). B.M. had found that Hindu couples often requested fetal sexing, wanting to terminate the pregnancy if the fetus was female; that there was a stigma attached to inherited diseases; that women often hid the fact that their child was unwell as much as possible, even from their own families; that women were blamed for bearing a sick child and bringing the disease into the husband's family; and that consequently many Hindu mothers of sick children were socially very isolated and often depressed.

Against the background of this experience B.M. initially expressed considerable surprise at the openness of the families, remarking, "They're such dignified people." I, as an insider, experienced no great surprise at the families' attitude and was rather taken aback at B.M.'s initial reaction, to which my reply was "But, of course, what were you expecting?" B.M.'s expectations, as explained to me, were as follows: Hindu people of Indian origin appear to have quite exceptional problems in coping with thalassaemia. British Pakistanis are likely to be poorer, less educated and more under the control of a doctrinaire religion. The problems for British Pakistanis would therefore be expected to be uniquely difficult - the extreme of underprivilege. This population had to be studied and the problems faced. The expectation was

that it would be a rather depressing experience. But the actual experience was quite different: the first thing B.M. noticed was the graciousness of manner of the couples and the equal participation of the women in the dialogue. This, for B.M., was not an isolated impression but was reinforced at every subsequent encounter.

We both perceived B.M.'s initial reactions as illustrating the different views of British Pakistani muslims held by Western society, and those of British Pakistani Muslims of themselves, and this led to continuing discussion. These and similar experiences, plus the findings of the research, made me dwell further on Western perceptions of British Pakistani family life, and particularly the status of the women in the family structure, as these expectations regarding submissiveness differed from my own and other British Pakistani Muslim women's (including those in the study) perceptions of themselves. The status of women in Islam or among British Pakistanis is not the concern of this thesis nor have I carried out an exhaustive theoretical analysis of the subject. However, the supportive manner and the strength with which women in this study dealt with their situation contrasted so starkly with popular Western perceptions of British Pakistani women that it warranted further consideration.

Popular preconceptions of British Pakistani Muslims in British society

Popular Western images of Muslims are that they are conservative in religion and often fanatic, examples of fanaticism being the rise of what is viewed as Islamic Fundamentalism in a number of Muslim countries, particularly Iran. In Britain this view has recently been fuelled by the events surrounding the Salman Rushdie controversy. The increase in Muslim political activity worldwide is viewed with apprehension, and dislike of what is generally thought to be a backward political, judicial and social system; the serious press recognise it as an unknown and alternative political and social force while the tabloids ridicule it (see Kabbani, 1989, Akhtar, 1989).

Within the above context British Pakistanis are generally viewed as belonging to a Muslim tradition that is authoritarian and grants males authority over females; that British Pakistani women are submissive and secluded in their homes and that their participation in public activity is scrutinized and controlled by males; that in all spheres final authority rests with the male members of the household.

However, a closer examination revealed that the strength the women displayed could be attributed to certain rights that Islam bestows on muslim women, of which both the men and women were aware. In addition, support structures for women as well

as men and children are inherent in the kinship pattern of societies that practice consanguineous marriage. These structures are in effect created by that marriage pattern. It is not within the scope of this thesis to investigate all the factors relating to what I found to be generally positive self-images among the women, only the factors relating to consanguinity are dealt with as this practice has come under particular attack, as the family studies have illustrated.

### Discussion

The consanguinity study, detailed earlier in the chapter, showed that 55% of marriages among British Pakistanis were to first cousins and 83% (including first cousins) were within the biradheri where both partners could trace a common ancestor. Shami and Zahida's study (1982) showed that amongst their sample of 900 in Lahore 36% of couples were first cousins whilst 73% were married within the biradheri. So most marriages among Pakistanis and British Pakistanis are within the extended family or within the biradheri where there is already a recognition of reciprocal family ties prior to a marriage being arranged. An individual growing up within this familiar framework already has individual and family ties of affection and responsibility with its members which are further consolidated through marriage. The introduction of a related in-law is always preceded or followed by the relationship before marriage, as in, "This is my cousin and also my sister/brother-in-law".

The family and biradheri networks are networks of affiliation and support but also networks of control. Therefore parents of a daughter arranging a marriage within the family or biradheri feel that their son-in-law and his immediate family, who the daughter generally lives with, remain answerable to them in terms of how their daughter is treated after marriage. The expectation is based not only on the ties forged through the marriage but also on the relationship that existed prior to it. This contrasts with societies where only exogamy is practised as the woman marries into a family where there was no previous link or shared life experience, as in most Hindu societies.

If the majority of the marriages within a society are in the same biradheri then it follows that the children of the marriages belong to the biradheri of their mother as well as their father. Hence females have as strong and supportive a network of relations as the males, encompassing the maternal and paternal sides of the family. It is because of this multiplicity of relationships (figs. 4.6 and 4.7) that most women are not isolated and they are not blamed for bringing a disease into the husband's family, for to blame one's wife's family is akin to blaming one's own family.

#### Attitudes towards consanguineous marriage

When families in the study and British Pakistani acquaintances of the researcher were questioned as to why they thought





consanguineous marriage was regarded favourably the following replies were given:

"We think it is comfortable."

"When a son or daughter marries in the family one knows the background of the family. They are the same as us".

"When you choose a neice or nephew there is already love and respect established with that person. There is the security that your son or daughter will be treated with respect. With outsiders, you never know what you're letting yourself in for. You have to be more careful".

"You have so much love in your heart for your daughters. You bring them up and you know that one day they will leave your home. So you try to arrange a marriage so that you can still be close to them".

"With your relatives, they know your financial situation. You don't feel you have to spend unnecessarily to maintain your status. They know the score".

"If a daughter dies, God forbid, then if the marriage is within the family, contact can still be maintained with her children".

"Sometimes it makes no difference whether you marry your relatives or not. Relatives can turn out to be worse than non-relatives. There's no guarantee in these things".

"We don't marry out of our quom. Neither the sons nor the daughters. There are some people who will marry the daughters of other quoms, because the children take on the quom of their father, but the women don't marry out".

When questioned about whether there was an overt economic motive in arranging marriages most people thought it

proposterous to even consider it stating that:

"Some people may consider their money but we don't."

One gentleman replied:

"It is only the greedy or the greedy and wealthy who think of such things."

In a society that practises a degree of segregation between the sexes, there is a marked difference between the relative informality in the family setting and the formality observed with outsiders of the opposite sex. Given this situation, parents have greater participation when their children make a consanguineous marriage, particularly in the life of their daughter, who traditionally lives separately with her husband or her husband and his family. However, if he is a relative, the residence is likely to be nearby and mother and daughter will continue to live daily in the same social circle.

With this small sample it is not possible to make definitive correlations, but the people who spoke less of quom affiliations were professionals of an urban background. Instead they emphasised being of the same educational background and being of a "good" family as important factors in considering a marriage partner. Explicit economic reasons for marriage were either not mentioned or regarded with distaste.

In general, regardless of background, people tended to say

that it was common sense to look for a marriage partner in their own inner circle and only move outwards if a suitable one is not available.

#### Adaptation of marriage patterns

The knowledge, prior to marriage, that one is at risk for having children with an inherited disease and the implications if one marries another carrier cannot be easily ignored. With this in mind prospectively counselled at-risk couples of Cypriot origin at the Perinatal Centre at U.C.H. were asked what they would have done if they had known they were both heterozygotes prior to their marriage. Some of those who had been introduced with a view to marriage said that if they had known from the beginning, they would probably not have married. However those whose marriage had been preceded by a romantic involvement said the information would not have affected their decision to marry (Modell and Berdoukas, 1984). It remains to be seen, and is an area of future study, which strategies British Pakistanis will employ to ensure healthy children, given their own cultural and religious context .

The existence of supportive networks for men, women and children seems to be one plausible explanation for the supportive manner in which British Pakistanis couples have dealt jointly with the care of their sick child and resolved the issues surrounding prenatal diagnosis. It also throws light on the reasons why British Pakistanis and Pakistanis as

a whole regard consanguineous marriage as a valuable social practice.

The above is not to suggest that every British Pakistani woman has a supportive structure around her, as not all marriages are consanguineous or will be in the future, and not all consanguineous marriages are harmonious. Hence, on an individual level, it should not be assumed that a couple necessarily operate within a supportive network. However, consanguineous marriage in most of the population sets a tone (standard) likely to be followed by non-consanguineous marriages.

THE CLINICAL IMPLICATIONS OF CONSANGUINEOUS MARRIAGE

"Consanguinity principally influences the category of disorders that are inherited as autosomal recessives". All other forms of inherited disease are largely unaffected by consanguinity (Harper, 1981).

Parental consanguinity in itself usually has very little impact on the offspring unless both parents carry a gene for the same recessive disorder. If the partners carry different disadvantageous genes, then consanguineous marriage has no major impact and therefore little genetic relevance for that couple. When it is known that there are genes for a recessive disorder in the the family (usually indicated by the birth of a homozygote) then it is possible for a geneticist to calculate the risk of having an affected child for two people who intend to marry within that family. The closer the relationship between the two people the greater would be the risk. The risk to a person of having affected offspring is only eliminated if he or she marries into another family that definitely does not carry the same recessive gene, or if they can be tested for carrier status and shown to be negative. If a non-related partner carries the same gene then the reproductive risks for the couple remain the same as for couples of carriers who are consanguineous. The solution therefore, does not lie in not marrying outside of one's family but in being tested for whether both marriage partners

carry the same disadvantageous gene. Some individuals who have consciously decided not to marry within their own family to avoid having an affected child, but still unknowingly selected a carrier partner and had an affected child, have been disappointed and felt let down by their medical advisers. The risk of having a child with thalassaemia major for people of Pakistani origin is not eliminated by "not marrying one's cousin" as was implied to the families in this study.

The impact of consanguinity, however, does differ according to whether heterozygotes for a particular disorder are rare or common within a given population.

#### Impact of consanguinity on the incidence of common recessive disorders

For a common condition, such as thalassaemia major (one in seventeen people of Pakistani origin are carriers), when a carrier chooses to marry into another family there is still a high chance that he or she may marry and reproduce an affected child. In Cyprus, where one in seven persons are carriers of the thalassaemia trait and there is no consanguineous marriage, at least among Greek Cypriots, there is still a high risk of marrying another carrier. The solution for both populations is the same: to test for carrier status and have an awareness of the the risks, if any, of having sick children.

Of the 4,000 known conditions with a Mendelian inheritance pattern, about one third are inherited as recessives. In this latter category carrier testing is possible only for the haemoglobinopathies (thalassemia and sickle cell disease), Tay-Sachs disease, and cystic fibrosis in the near future. In the absence of carrier testing, reliance has to be placed on the calculation of risk, hence, for common conditions there can be no guarantee of escaping a recessively inherited disease merely by marrying out.

Impact of consanguinity on rare recessive disorders

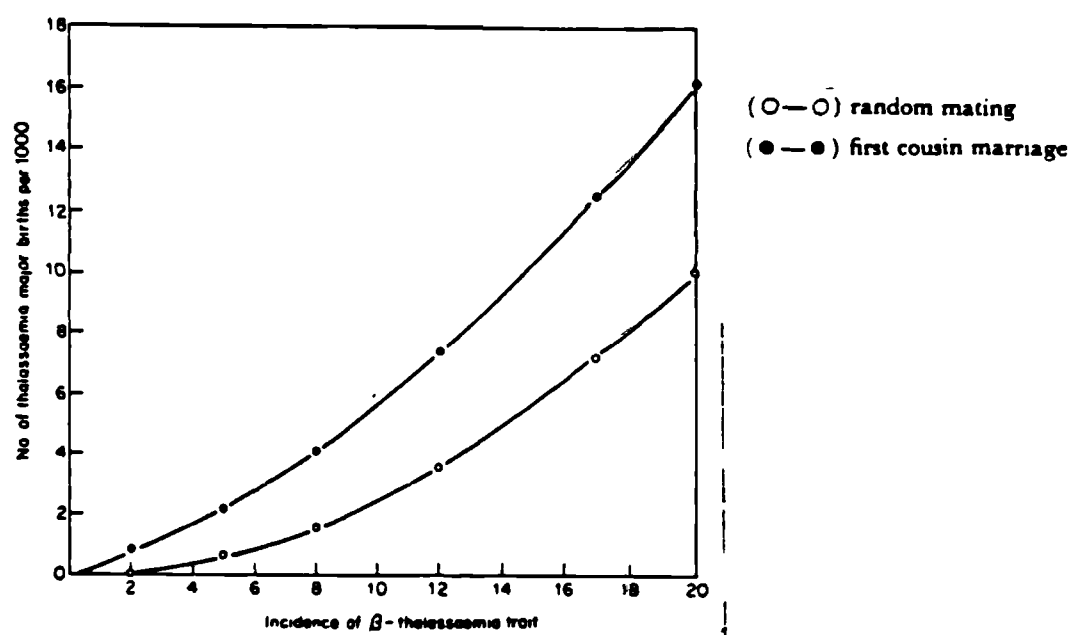


Fig. 4.9<sup>6</sup>: Relationship between heterozygote prevalence and homozygote birth.

As the above figure shows, the impact of first cousin marriage

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<sup>6</sup> Source: W.H.O., (Milan and Bangkok, 1985). Model educational aids to assist in haemoglobinopathy control. p.8.10, HMG/G/85.8a.

for rare recessive conditions (i.e., carrier frequency is 1% of the population or less) is much greater. If a rare recessive condition has manifested itself in a family then the chances of the same gene being in another family is far more remote than it being in the same family. When a rare condition has already been identified in the family by the birth of a homozygote, marrying a non-related person would considerably reduce the risk of having an affected child.

It is often thought that a high frequency of consanguineous marriage explains a high frequency of inherited disorders, including haemoglobinopathy traits. In fact, though a consanguineous marriage pattern increases the frequency with which the recessive mutant genes present in the population are expressed in terms of leading to the birth of a homozygote, with the passage of time the custom operates to decrease the frequency of abnormal recessively-inherited genes in the population since with every homozygote born who does not reproduce, two genes leave the population (Bittles, 1980). There is evidence that this has happened in parts of Southern India where close consanguineous marriage has existed for several thousand years.

Regardless of the frequency of recessive lethal genes, in populations where consanguineous marriage is common most affected children are found among the offspring of such marriages, and tend to be clustered in family groups rather



than scattered randomly. However, meticulous genetic epidemiological work is needed before it can be stated with confidence that the general frequency of recessively inherited conditions is, or is not, unusually elevated in a given population.

Regardless of whether a condition is common or rare, as Roberts states:

"Once a consanguineous couple has produced a child with a recessive defect, the risk of its recurrence is virtually the same (1 in 4) as for any other parents of a child with this condition, though there is an increased risk of some other defect. It is doubtful whether in counselling the fact that the parents are consanguineous need even be mentioned. If one is asked whether this tragedy is due to their relationship, one can only say that this has contributed, but point out that if they had both chosen other partners they might have produced children with some other condition, for everybody carries bad genes for some disorder or other" (Bundey and Roberts (1988), p.35).

To discourage consanguinity without an analysis of an individual's particular situation is to offer advice which has no factual base. It is both unnecessary and harmful. Commenting on this issue Harper (1981) states that:

"The moral for the clinician should be to beware of generalisations, and to realize that in the majority of situations the advice given to individual couples may have a profound effect on them and their offspring, but will rarely alter the population structure to a significant extent (p.268)."

The differing patterns of manifestation of recessively inherited disorders

The study of a recessively inherited disorder in a population that practices close consanguineous marriage provides a valuable "tool" for identifying at-risk families. Due to the consanguinity, recessively inherited conditions tend to be clustered in family groups rather than scattered at random throughout the population, as would be found in a population with a non-consanguineous marriage pattern. The rarer the gene, the higher will be the proportion of parents who are consanguineous. The commoner the gene the higher will be the proportion of non-consanguineous parents (Figs. 4.7 and 4.8).

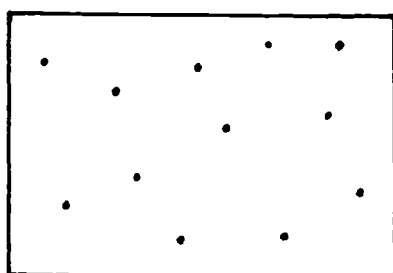


Fig. 4.10: The manifestation of a recessively inherited disorder in a population not practising first cousin marriage.

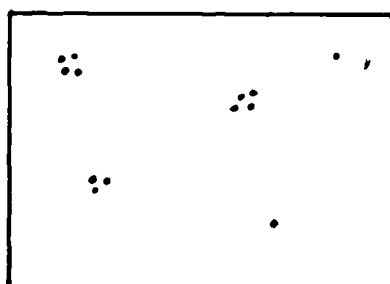


Fig. 4.11: The manifestation of a recessively inherited disorder in a population practising first cousin marriage.

The birth of an infant with a recessively inherited disorder identifies a family that is transmitting the gene. When consanguineous marriage is common and family size is large, as in the British Pakistani population, most recessively inherited conditions will mark their presence by leading to the birth of an affected child somewhere within the cluster. Genetic diagnosis and counselling focussed on the family of affected infants may then be particularly effective in detecting couples at risk and advising them prospectively, that is, before the birth of an affected child.

In Britain, populations with different marriage conventions and the consequent differing patterns of manifestation of inherited conditions live side-by-side, with one health service responsible for the delivery of genetics service to the whole population. In North-West Europe, including the UK, there has so far been limited success in providing a genetics service for disorders that affect diverse and unevenly distributed ethnic minorities (Royal College of Physicians, 1989, p.26). However, in reality the specific pattern of manifestation of recessively inherited diseases among British Pakistanis can be used to advantage in developing and delivering a highly effective and sensitive genetics service to this group of people, with some adaptation of the present service.

CONCLUSION: A POSSIBLE APPROACH TO GENETIC COUNSELLING AMONG  
THE BRITISH PAKISTANI POPULATION.

Using each family with a homozygote as a starting point, the genetic counsellor responsible for seeing the families, is able to counsel the family of the homozygote and sensitively move outwards to the siblings of the parents and so on, and so provide counselling for a cluster of families with a known genetic risk. This study has already shown that parents discuss the condition openly with other parents of affected children and within their own family. The family studies illustrated situations where the condition was discussed among extended family members and some said that, as the condition was inherited and could affect other family members there was a need for them to know. Some parents asked if there was any written material they could pass on to other people. Mrs. G. wished the researcher to approach her brother and his wife tactfully to explain the inherited nature of the condition, so that they would have their children tested and she could then ask for the hand of one of their non-carrier daughters for her carrier son. She felt the impetus to test their children would be better coming from her brother and sister-in-law than through any pressure from herself. She also felt the information would be far better received from an informed, neutral outsider than from within the family. In general, couples felt they could discuss the condition with their siblings but did not feel they could or should pressurise any siblings to be tested. The role of an informed outsider in

providing accurate information was recognised and felt to be important.

Alongside the work with family clusters the establishment of support groups could bring together the various families with already affected children, creating networks between families where information and support could flow (Fig. 4.9).

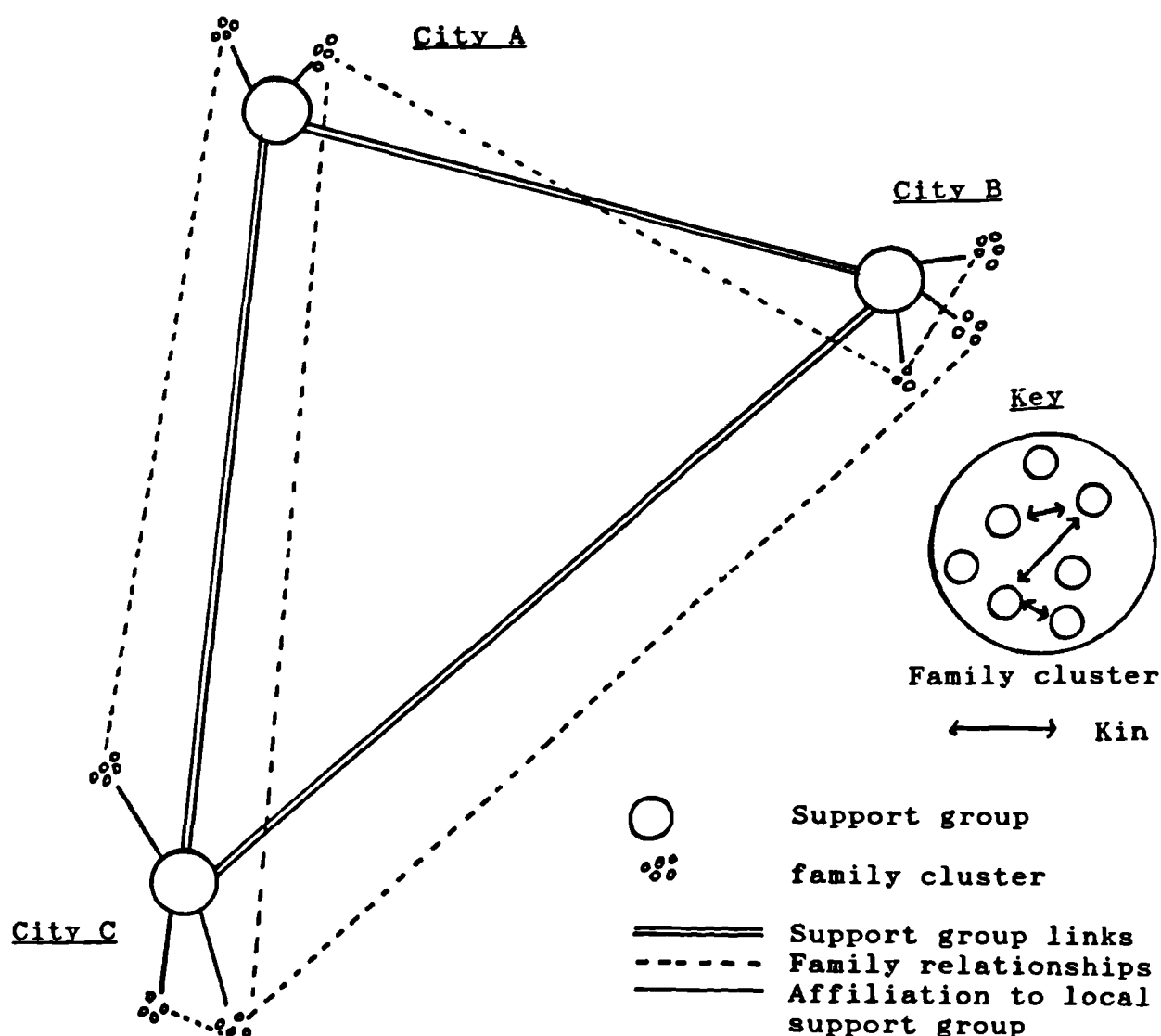


Fig. 4.12: Model showing possible networks of information and support formed through genetic counselling and support groups.

The kinship and marriage patterns among British Pakistanis are such that networks of kin exist in neighbourhoods, in cities and between cities. In the duration and limitations of this research project, it was found that families in other parts of England had become aware of the developments in city Z. and the adjoining areas through informal family networks. I myself was contacted directly by families, unconnected with the research, from two distant cities. Visits also took place between distant kin or unrelated couples with affected children within city Z. and between different cities to discuss experiences of bone marrow transplantation and prenatal diagnosis. Sometimes friends and relatives of the family also attended the support group meetings.

The willingness of British Pakistanis to discuss and share their experiences and to offer support does not mean that inter-familial or inter-communal conflicts do not exist. It is important that the counsellor establishes a good rapport with each family, is aware of the nuances of community and family life and discusses the possibility of working with the extended family with the couple first.

For the detection of heterozygotes scattered randomly in the community the same services available to the rest of the population should be operational, i.e., routine screening at antenatal clinics, G.P. surgeries, Well Woman clinics and so on. It is, however, necessary to assess whether these

mainstream channels are actually being used effectively by minority populations: if not, then which modifications and initiatives are required? It is only by a comprehensive and integrated approach to the development of appropriate health services that any degree of success can be achieved.

As rare conditions are even more likely to be limited to family clusters, the same approach could be applied to families with other recessively inherited diseases and not just to thalassaemia. In this study two of the families were also transmitting the gene for cystic fibrosis and one family was also transmitting maple syrup urine disease.

The proposed approach to counselling and imparting information to families is non-intrusive, as it begins with dealing with families who are in greatest need of the genetics service, in their homes which is their territory, and in a cultural context which is their own. By focussing on the area of need and diffusing outwards rather than on the community as a whole and working inwards, it is also non-alarmist. The proposed strategy avoids the time, expenditure and pitfalls of a health education campaign trying to impart complex genetic information, a scanty knowledge of which may do more harm than good. In Britain, where racial tensions already exist, it is important for health care initiatives not to single out and advertise that minority groups have health problems peculiar to themselves, especially when they elicit prejudiced

reactions as consanguinity does. Information on an inherited condition should always be presented in the wider context of different genetic conditions affecting every population worldwide. Until it is a widely known and well established fact that all populations have to contend with inherited diseases, any initiative in genetic screening and counselling should be treated with extra care and sensitivity.



### FINAL CONCLUSION

The limitations of present ad hoc facilities for screening for haemoglobinopathies (see Prashar et al, 1985) have caused much distress; not to screen is inequitable but to screen without counselling can do more harm than good. An integrated and comprehensive approach to screening with adequate and appropriate genetic counselling facilities is essential. As the genetics service is still in its infancy, a recent report of the Royal College of Physicians has addressed this specific area (Report of Royal College of Physicians, 1989). In fact the findings of this study contributed to development of some of the concepts proposed in the report. Their recommendations include a model for a community genetic screening service (Fig. 4.10).

The role of the specialist trained genetic counsellor<sup>7</sup> (A), remains tentative at the moment (B. Modell, personal communication) but it is envisaged that existing non-medically trained specialists<sup>8</sup> all of whom have experience of relating to the community and community structures, and who are also more in touch with primary care than highly specialised doctors are required. They should be further trained in the broader aspects of genetics and help to develop a regional

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<sup>7</sup> The term "specialist genetic counsellor" is still under review, as it is felt it does not fully encompass the range of responsibilities of this senior post.

<sup>8</sup> Non-medically trained specialists who are already developing and delivering specific services, e.g., for haemoglobin disorders or working with clinical geneticists.



genetics service in collaboration with the specialists indicated in circle B, taking into account the ethnic diversity of the population. The report also recommended that each region should set up a management group responsible for the delivery of the service.

For regions with a high concentration of British Pakistanis it is essential that the specialist genetic counsellor, whose responsibilities would include a specific service for British Pakistanis, should be a female of the appropriate religious, linguistic and cultural background.

The role of the specialist counsellor should include (1) research, (2) development, (3) training and (4) monitoring components. The range of methods used by the specialist counsellors to develop a comprehensive service will undoubtedly develop over time once the counsellors are in post. Examples of the above could be the training of health professionals (G.P.s, health visitors, midwives, socialworkers etc.) about the applicability of their roles in the genetics field and the nuances of working with British Pakistani families; obtaining funding for further genetic counsellors to be specifically trained to counsel British Pakistani families and run support groups in the region; and the development of appropriate health education materials.

Bringing together the conclusions of the family experience of thalassaemia (part 3, pp. 261-263) and consanguinity - social and health implications (part 4, pp. 297-301), leads to the final conclusion that the role of the specialist counsellor, discussed above, is the starting point to delivering an adequate and appropriate service for British Pakistanis.

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APPENDIX 1<sup>1</sup>: The distribution of British Pakistanis in BritainRegion

	<u>Total (NCWP)</u> (%)	<u>Pakistan</u> (%)
<u>Scotland</u>	46,188 (2.1)	9,903 (3.4)
Central Clydeside	21,595 (1.0)	6,722 (2.3)
Remainder of Scotland	24,593 (1.1)	3,131 (1.1)
<u>England</u>	2,136,590 (96.8)	282,329 (95.6)
<u>Wales</u>	24,467 (1.1)	3,229 (1.1)
<u>North</u>	26,247 (1.2)	6,343 (2.1)
Tyne & Wear Met. County	11,367 (0.5)	2,132 (0.7)
Remainder of North	14,880 (0.7)	4,211 (1.4)
<u>Yorkshire &amp; Humberside</u>	154,344 (7.0)	60,215 (20.4)
South Yorkshire Met. County	24,606 (1.1)	8,947 (3.0)
West Yorkshire Met. County	118,372 (5.4)	50,556 (17.7)
Remainder of Yorks./H.	11,366 (0.5)	712 (0.2)
<u>East Midlands</u>	140,991 (6.4)	11,906 (4.0)
<u>East Anglia</u>	28,429 (1.3)	3,856 (1.3)
<u>South East</u>	1,227,754 (55.6)	88,811 (30.1)
Greater London	954,148 (42.8)	52,192 (17.7)
Outer Metropolitan area	186,459 (8.4)	28,987 (9.8)
Outer South East	96,147 (4.4)	7,632 (2.6)
<u>South West</u>	63,607 (2.9)	4,446 (1.5)
<u>West midlands</u>	326,523 (14.8)	61,931 (21.0)
West Midlands Met county (18.6)	285,350 (12.9)	54,819
Remainder of West Midlands	41,173 (1.9)	7,112 (2.4)
<u>North West</u>	168,695 (7.6)	44,821

<sup>1</sup> Source: OPCS Census 1981. In Anwar, M. ( 1985), pp. 251.

Greater Manchester MC	100,045 (4.5)	(15.2) 25,572 (8.7)
Merseyside		
Met. County	14,823 (0.7)	727 (0.2)
Remainder of		
North West	53,827 (.4)	18,522 (6.3)
 Total (100)	 2,207,245 (100)	 295,461